LETTERS TO THE EDITOR

Cystic fibrosis identified by neonatal screening: incidence, genotype, and early natural history

EDITOR,—Commenting on our paper, which described an apparent halving in the incidence of cystic fibrosis in East Anglia,¹ Dr Coles and colleagues suggest that during the last three years of the reported study, a substantial number of patients with cystic fibrosis might have remained undiagnosed because of over reliance on the efficiency of the screening test.² This is an implausible explanation for our observations. Such an occurrence would require either a drastic deterioration in the analytical performance of the assay, which is not supported by the results of the external or internal quality control procedures conducted throughout this period, or an abrupt change in the pathophysiology responsible for the raised blood immune reactive trypsin (IRT) concentrations in the newborn who have inherited cystic fibrosis during this later period. Furthermore, there is no evidence to suggest that East Anglia's paediatricians have, over the years, become complacent about achieving the earliest possible diagnosis in that small percentage of cystic fibrosis patients not detected by screening. We are confident in this assertion from the number of inquiries received from every ward and clinic in the region seeking to confirm that routine neonatal IRT screening has been carried out on infants and children presenting with symptoms, in conjunction with the continuing usage of the excellent sweat testing facilities throughout the region, and more recently with the increasing use of the facility to test for the more common cystic fibrosis genotypes on DNA extracted from stored neonatal blood spots.

Secondly, all the cases were diagnosed by IRT assay. Dr Coles and colleagues refer to the figure of 68% of cases being identified by IRT assay alone. The remainder of cases had clinical features which might have led to the diagnosis, in addition to an abnormal IRT assay.

There is thus no evidence to suggest that under ascertainment explains the declining incidence. We agree that it will be important to continue monitoring the incidence of cystic fibrosis in East Anglia.

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1 Green MR, Weaver LT, Heeley AF, et al. Cystic fibrosis identified by neonatal screening: incidence, genotype and early natural history. *Arch Dis Child* 1993; 68: 464-7.

2 Coles EC, Dodge JA, Morison S. Cystic fibrosis identified by neonatal screening: incidence, genotype and early natural history (letter). *Arch Dis Child* 1993; 69: 470.

Parental participation in case conferences

EDITOR,—In child protection work the focus of professional endeavour should be the welfare of the child and as children live (usually) in families, their carers must clearly be involved in any assessment and longer term plans. So what is the function of the child protection conference? In the past information was collected concerning possible abuse and an assessment of the child and family at least started. Now the conference has altered, it is held at the end of an investigation usually jointly planned by the police and social services and reports are focused and tend to look at the incident rather than attempt to look at the concerns in the context of the family. The presentation may be further modified by previous strategy meetings where the professionals have met and discussed in some depth the gathered information. It is then the task of the case conference to consider whether inclusion on the child protection register is to be recommended, a key worker and core group is identified.

The parents will usually work with the various professionals and their attendance at the conference is a logical extension of this work. But what of the difficult cases where parents strongly deny the concerns and do not accept the need for professional involvement? As colleagues increasingly recognise child sexual abuse they will meet more denial. A 3 year old girl told her mother of the 'games' she plays with Daddy on her weekend access. Social services and the police investigate, the medical reveals a torn hymen and Daddy denies any wrong doing. Although the professionals feel there is substance in the story the Crown Prosecution Service do not take the case forward. At the conference are the estranged parents, the mother may have worked closely with the professionals, the father asserts it's all an attempt by his malicious ex-wife to stop access. Do we have the skills to handle all the issues presented here in the conference, attended by the parents' two solicitors who may advise their client not to talk?

It may be argued that the above case is unusual: this depends upon the geographical area and the willingness of professionals to engage in complex work. Each is different, if the girl in the scenario above was 13 years would she want her father, who may be her abuser, to have details of her medical report (and her teacher, etc ...).

Working Together does not expect that professionals can always work with parents to achieve child protection.¹ Cooperation may develop but true cooperative 'partnership' is not achieved instantly. A working relationship with parents may be a better description of the longer term expectation.

In recent papers in the journal *Sheehy Skeffington* is right to suggest caution and keep the focus on the child's needs and welfare²; Hutchinson is overly optimistic of parenting behaviour given our increasing knowledge of the dynamics of abusive relationships and behaviours.³

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1 Home Office. *Working together under the Children Act 1989*. London: HMSO, 1991.

2 Sheehy Skeffington F. Parental participation in case conferences: the case against. *Arch Dis Child* 1993; 69: 457-8.

3 Hutchinson T. Parental participation in case conferences: the case in favour. *Arch Dis Child* 1993; 69: 455-7.

EDITOR,—We write in full support of parent participation at case conferences as presented by Dr Hutchinson.¹ We would however like to draw attention to some recent problems.

On one occasion the parents were accompanied by all four grandparents all wanting to voice their opinions against the medical diagnosis of non-accidental injury and give character references of the parents. Chairing such a conference proved extremely difficult. One parent appeared inhibited by their presence and only went on to give relevant information after the conference ceased and the group dispersed.

At another case conference a solicitor accompanied the parent and took copious notes using the occasion to 'interrogate' the doctor on the significance of her medical findings in a 'court trial format'; the conference lasted five hours.

On another occasion a female friend accompanied the mother for support. Her surname (given on introduction to the conference) was the same as the alleged perpetrator's alias, although she claimed not to know him. She was, in fact, his sister and after the conference was in a position to give him some valuable information regarding the investigation. The health visitor was clearly anxious about stating her concerns for the family openly. The mother without warning suddenly lost her temper and attempted to physically assault the health visitor who was left extremely shaken by the incident and later had time off work.

We recommend that a time limit is imposed on conferences. The number of supporters should be limited. They should be there purely in a support rather than a contributory role. Information on them should be checked before the sharing of such confidential information especially if an investigation is still proceeding.

It is our general experience, however, that parental participation promotes partnership and good practice and should continue.

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1 Hutchinson T. Parental participation in case conferences: the case in favour. *Arch Dis Child* 1993; 69: 455-7.

Sudden infant death syndrome

EDITOR,—A high degree of heat production has been identified as a risk factor in the causation of sudden infant death syndrome (SIDS) by several studies.¹ It has been suggested that one reason why sleeping prone may increase the risk of SIDS is that it restricts the loss of heat from the infant to the environment.¹ Concern has thus focused on factors that may increase heat production, such as febrile illness, or limit heat loss, such as excessive thermal insulation of clothing or high environmental

temperature.¹ We suggest that mode of feeding be added to the list of factors being investigated with regard to increased heat production.

Differences between breast fed and formula fed infants in metabolic rate, which is directly related to heat production, have already been reported,² and we have recently found sleeping metabolic rate to be significantly higher in formula fed compared with breast fed infants at age 12 weeks. Furthermore, total daily energy expenditure (TDEE) measured by isotopic methods has also been shown to be significantly greater in formula fed than breast fed infants.^{2,3} In a longitudinal study in the first year of life, TDEE was found to be significantly greater in formula fed infants at 6 weeks and 12 weeks but to be similar between the diet groups at 6 months and 9 months (P S W Davies, unpublished data). This pattern correlates with the reported distribution of SIDS by age.

A number of studies of SIDS have reported higher rates of formula feeding in cases compared with controls.^{4,5} Because SIDS does occur in breast fed infants, formula feeding has not been considered as a major risk factor. Some authors have suggested that the relationship between formula feeding and SIDS incidence is an artefact of the relationship between SIDS and social class.⁵ However, it is more likely that the reverse is true, and that variables such as family size, social class, maternal age and interpregnancy gap are related to SIDS incidence because of their effect on aspects of infant care, of which formula feeding might be one.

If this hypothesis is correct, one explanation might be that the amount of energy in contemporary formulas is high in comparison to the mean energy content of breast milk. This view has been increasingly supported from recent studies of nutrition, growth, morbidity, and development of breast fed and formula fed infants.^{6,7} These findings support the need for a thorough review of energy requirements in infancy and especially the adequacy of the energy density of infant formulas.

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- 1 Fleming PJ, Gilbert R, Azaz Y, *et al.* Interaction between bedding and sleeping position in the sudden infant death syndrome: a population based case-control study. *BMJ* 1990; 301: 85-9.
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- 7 Garza C, Butte NF. Energy intakes of human milk-fed infants during the first year. *J Pediatr* 1990; 117: S124-31.

The physician's hands and early detection of neuroblastoma

EDITOR.—The data from some developed countries show that within the past 25 years, the five year survival rates in neuroblastoma have increased twofold from the initial 25%.^{1,2} The outlook for patients presenting over 1 year of age with stage IV remains dismal (20% five year survival). The drop in mortality rates is related to more frequent detection of neuroblastoma in children before age 1 and before their disease is advanced¹ as well as more frequent incidental detection of the neoplasm³⁻⁶ (see also Carlsen⁴ for further references).

In the years 1943-80 in Denmark, the percentage of incidentally detected neuroblastoma increased from zero to 14%. Among children with incidentally detected neuroblastoma, stages I and II predominated (16 out of 20 cases). The total number of children treated for neuroblastoma was 250, and nearly one half of all 53 long term survivors were found incidentally or had 'spontaneously regressing' tumours.⁶ In Germany scrupulous medical examinations of children, especially below 1 year of age, during obligatory frequent check up visits to which almost 90% of parents faithfully report, allowed the incidental detection of neuroblastoma in every sixth patient with this tumour. Out of 65 children at stage I and 60 patients at stage II, incidental detection of neuroblastoma occurred, respectively, in almost every second and third patient with this disease.⁵ This may suggest that a systematic approach,⁵ a greater awareness of the relatively high incidence of this tumour (the most common solid tumour in children), and the need for good abdominal examination may increase the detection of children with neuroblastoma before the onset of symptoms.^{5,6} The shift to diagnosis at earlier ages and stages may result from more frequent chest radiographs and use of ultrasonography of the abdomen.⁵ Sawada *et al* found that even a small abdominal tumour of neuroblastoma can be detected by careful examination. Out of 293 infants suspected of neuroblastoma on the basis of urinary screening, physical examinations revealed a tumour in more than one half of the patients.⁷ A careful abdominal examination is of a great importance in neuroblastoma (the primary tumour is in 75-95% of the cases located within the abdomen).

The hands and eyes of a physician have always been and continue to be the most important tools in detecting diseases. A physician may notice slight, rare symptoms of low stages, strongly suggestive of a neuroblastoma, such as Horner's syndrome and associated heterochromia, the watery diarrhoea syndrome and the dancing eyes and dancing feet syndrome.

Never in his entire life is a human being subjected to medical examinations as often as in early childhood. This is dictated by obligatory periodic check ups, physical examinations before vaccinations, and a mother's loving care, prompting her to seek medical assistance any time she sees a sign or symptom which makes her anxious. The skilful hands of a physician, carefully examining the abdomen on these occasions, can contribute to early detection of the low stages of neuro-

blastoma⁴⁻⁵ and to the decrease of mortality rates in this tumour. It is possible to perform this type of screening throughout the world.

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- 1 Murphy SB, Cohn SL, Craft AW, *et al.* Do children benefit from mass screening for neuroblastoma? Consensus statement from the American Cancer Society workshop on neuroblastoma screening. *Lancet* 1991; 337: 344-6.
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SPRING BOOKS

Managing Children with Psychiatric Problems. Edited by M Elena Garralda. (Pp 212; £14.95 paperback.) BMJ Publishing Group, 1993. ISBN 0-7279-0788-3.

At last - a book about my craft which I can identify with and recommend to trainees and others who may be wondering what we actually do. For me this book fills a gap, amplifying those textbooks that have focused on describing the various conditions or predicaments which we deal with as child and adolescent psychiatrists. The aim of this book is to help paediatricians and other doctors address the psychiatric aspects of children's health problems. The editor, Professor Elena Garralda, adds that she hopes this book will be of interest to not only doctors but also teachers, social workers, and to our own psychiatric trainees. She also hopes that the book will help in the referral of disturbed children to specialist services. The contributions have all been reprinted from the series in the *Archives* entitled 'Types of Psychiatric Treatment', and which ran for 14 issues.

The first two contributions deal with the identification of psychiatric disorders in children followed by a brief overview of the types of available psychiatric treatment, as well as the all important question of efficacy. The remainder of the book then amplifies the types of treatment and management approaches which we use. I cannot pick out one or two chapters for special mention, which is an indication of the high level of each of the