LIVER CIRRHOSIS IN CYSTIC FIBROSIS

EDITORS.—Feigelson et al report a relatively large series of 31 patients with cystic fibrosis and liver cirrhosis, emphasizing its protracted course and reviewing their experience in the management of portal hypertension.1 We too have observed that the development of uncomplicated cirrhosis often has little influence on the natural history of cystic fibrosis. However, we disagree with their conclusions about the management of alimentary bleeding from portal hypertension, which is a life threatening complication. In their study, five patients were treated by injection sclerotherapy, which was limited to two or three sessions. Not surprisingly, recurrent bleeding was common and this, combined with four deaths from variceal haemorrhage and the potential for ectopic variceal bleeding, prompted the authors to express doubts about this technique. Their single success with partial splenectomy led them to conclude that this is the procedure of choice.

Since 1979 we have regularly reviewed 19 patients with cystic fibrosis, cirrhosis, and portal hypertension (figure), 11 of whom were included in a previous analysis of the results of injection sclerotherapy in children with bleeding oesophageal varices.2 Fifteen of these patients (eight boys, seven girls) have required injection sclerotherapy for variceal bleeding. The mean age at onset of bleeding was 11.5 (range 6–14.9) years. Variceal obliteration was achieved after a mean of 6 (range 3–9) injection sessions over a period of 0.8 (0.3–2.5) years; one boy is still undergoing treatment.

Five of the 15 died of respiratory failure without further gastrointestinal bleeding two months to nine years after variceal obliteration. Two patients have required surgery; one a splenectomy for symptomatic hypersplenism and the other a splenectomy and lienorenal shunt for gastric variceal bleeding. Excluding the boy still undergoing treatment, none of the survivors have significant residual oesophageal varices after a mean follow up of 4.5 years after completing treatment. Endoscopic injection sclerotherapy has proved effective in the control of oesophageal variceal bleeding and only one patient has required portosystemic shunt surgery for ectopic variceal haemorrhage. Feigelson et al report no respiratory problems from sclerotherapy. We have a policy of intensive perioperative physiotherapy, bronchodilators, and prophylactic antibiotics to minimise the recognised deleterious effects of general anaesthesia and sclerosant injection on respiratory function.3

Sclerotherapy is an effective and safe modality for the control of bleeding from oesophageal varices in children with cystic fibrosis and the results appear to be better than those of portosystemic shunt surgery.4 Nevertheless, such surgery has an invaluable role in the management of some patients, such as those with ectopic variceal bleeding.5

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Hairst loss in children

EDITORS.—We have read with interest the article by Verbóv on hair loss in children. Alopecia is a frequent manifestation of biotin dependent multicarboxylase deficiencies: while one of these disorders, holocarboxylase synthetase deficiency is very rare, that of biotinidase deficiency is more common with an estimated frequency of 1:84 000 world wide.2 Biotinidase is responsible for the processing and recycling of biotin from biotinyl peptides. Development of symptoms in biotinidase deficiency is gradual, with episodes of remission perhaps modulated by the availability of free biotin in the diet. Neurological manifestations are the most frequent initial symptoms but patients have also presented with primarily respiratory symptoms. Alopecia frequently occurs, and is a hallmark of the disease, with discrete hair loss that in some cases becomes complete.


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Cases have been mislabelled as acrodermatitis enteropathica on occasion. Biochemical indicators, for example an abnormal organic aciduria, are not always present and thus the observed symptoms become most important in the differential diagnosis of such patients, particularly with neurological and dermatological problems. Irreversible brain damage and even death may occur in untreated patients and as the disorder is rapidly responsive to oral biotin, early diagnosis and treatment is essential. Diagnosis is straightforward, by biotinidase activity determination in plasma or serum, and the condition should be considered in any cases of hair loss particularly associated with neurological, dermatological, or respiratory illness.

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Chicken scratches

EDITOR – Non-fatal self inflicted intended harm is commonly called ‘attempted suicide’ or parasuicide. The rate of parasuicide was reported to be declining in the 1980s when compared with the 1970s. In the last two decades there has been an increase in the rate of suicide in older children. The full extent of parasuicide behaviour in teenagers is not clear and a great proportion of parasuicides do not come to the notice of official agencies. Clinical studies on victims of child sexual abuse report that there is a higher than normal association with parasuicide. The youngsters do attempt suicide methods that are more likely to be discovered.

‘Chicken scratches’ is a new term used by teenage girls for the lacerations and bruising on the dorsal aspect of one or both hands (figure). These are produced by the vigorous rubbing of a plastic ruler. Teenagers are usually secretive, protect their peers’ interest, and so it is difficult to know how these injuries are inflicted and why they are seeking the attention of others.

It is important to identify these very obvious lacerations, which are not confined to the location of any major blood vessels, and could be dismissed as accidental injury. They are common in girls who are withdrawn and not popular in their class. These girls do not have friends or adults with whom they can talk about their own problems and worries. Once they are identified it will be necessary to involve the child protection agencies early on to prevent disastrous consequences.

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BOOK REVIEW


This book provides an interesting blend of good sense, some equivocal recommendations on management of health associated problems, and more than a whiff of ‘jolly hockey sticks’. It is likely to prove more use to doctors attached to independent schools, particularly those with boarders (presumably the bulk of Medical Officers of Schools Association (MOSA) membership) than to a clinical medical officer or rotating senior house officer attached to a state primary or comprehensive school. This is the 17th edition, the first was published in 1885, again in 1889. Some of the phrases used seems more applicable to the 1890s than the 1990s.

The book contains sections on the responsibilities of school medical officers, preventive medicine in schools including in the various physical and emotional development in childhood and adolescence, selected medical problems, sections on children with special needs, school child abuse, sports injuries, safety at school and the largest section, 47 pages, on communicable diseases including AIDS and exotica such as hepatitis E and schistosomiasis. There are eight appendices including lists of notifiable diseases, suggested entry questionnaires, advice to parents of boarding school pupils, development and growth charts, the Education ‘School Premises’ Regulations 1981, and swimming pool disinfectants.

Most of the advice is sensible and orthodox, if in some sections brief to the point of being incomplete. The easy to use layout will ensure likely frequent use of the book by MOSA members. The sections on alcohol, tobacco and misuse, especially the latter, are not optimistic regarding prevention and not really helpful regarding established use. The section on sex includes a good summary of acceptable procedures regarding prescription of contraceptives for girls, and includes the perhaps unintentional restrictive advice that ‘lodges taking the contraceptive pill should not smoke’.

There are only six pages on children with special needs including one on gifted children and three on children from ethnic minorities. The section on school child abuse seems to mainly relate to child sexual abuse but fails to distinguish acute from chronic, nor make the distinction into low, medium, and high probability. There are other concerns in this section, for example the social services or NSPCC are recommended to be engaged at the onset apparently only in the most blatant examples of school child abuse. When the history is equivocal, it is considered preferable for the physician to ‘maintain a low profile to avoid unnecessary anguish and distress for all parties’.

This book may prove useful, with reservations, to its organisational membership, but is likely to be of only limited use to most school doctors in this country and even fewer school nurses who, increasingly, provide health service continuity in schools as educational medicine moves towards becoming mainly a special needs service.

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