Recurrent cyanotic episodes with severe arterial hypoxaemia and intrapulmonary shunting: a mechanism for sudden death

Sir,—In a recent paper Dr Southall et al suggest that rapid shunting of un oxygenated blood through the pulmonary vascular bed is a cause of sudden and unexpected death in infants and children and that this may be causally related to the sudden infant death syndrome (SIDS).¹ This assertion is based on the occurrence of sudden death in eight of their 51 patients (16%), four of whom died during cyanotic episodes.

While the hypothesis and the physiological evidence for it are interesting and well presented, we note that the fatal cases reported that five of the eight had concurrent pathological processes that are independently associated with sudden death, and in some cases they had cyanosis: intact intrapulmonary shunts of blood in a patient with a repaired tracheoesophageal fistula (n=1), bronchopneumonia (n=2), bronchopneumonia with mucus within airways (n=1) and a brainstem glioma (n=1).² It would strengthen the underlying hypothesis considerably if further information could be provided detailing how these well established causes of sudden death were overshadowed by the intrapulmonary shunting as the prime mechanism responsible for death. Of even more importance is the need for clear documentation of the evidence for intrapulmonary shunting in the eight fatal cases, if this is to be accepted as the cause of death. It appears from the text that only one patient had contrast echocardiography (case 18) and that none had krypton infusion scans or postmortem injection studies. It would clarify their case considerably, therefore, if the antemortem and postmortem evidence for shunting in each of the fatal cases could be provided.

ROGER W BYARD
Department of Histopathology, Adelaide Children's Hospital, North Adelaide SA 5006, Australia


Dr Samuels and Southall comment: Dr Byard has raised important questions concerning our hypothesis that severe hypoxaemic episodes may cause sudden death in infants and young children. The presence of a structural abnormality at postmortem examination does not in itself always explain the cause of death. The cessation of cardiorespiratory and cerebral function must ultimately encompass a physiological mechanism. In our patients, the findings of bronchopneumonia, an intrapulmonary shunted impacted food bolus, and a brainstem glioma were alone insufficient, in our opinion, to explain their sudden and unexpected deaths. Two died during the night without any premonitory symptoms or signs, while the other three died during typical cyanotic episodes.

In all eight cases, our patients had suffered repeatedly from sudden and life threatening cyanotic episodes needing resuscitation. In the absence of physiological recordings during their deaths, we can only speculate as to the mechanism which caused their death based on the investigation of living infants undergoing similar cyanotic episodes and noting characterising characteristics that could be described as life threatening. Although some of our patients did not undergo all the investigations required to confirm the development of an intrapulmonary shunt during their cyanotic episodes, their episodes were identical in all other respects to those occurring during krypton infusion scans or contrast echocardiography. Intrapulmonary shunting is one of the best explanations for the sudden development of their life threatening hypoxaemia. How and where in the lungs this shunt occurs remains to be elucidated.

At present, the explanation for the final pathophysiological pathways leading to sudden and unexpected infant deaths, even in the presence of abnormal postmortem findings, remains unresolved.

Measles immunisation

Sir,—Active immunisation is efficient in preventing the central nervous system complications of measles.¹ The wisdom of current recommendations² on measles immunisation is illustrated by a case of progressive measles encephalitis after renal transplantation for treatment of the congenital nephrotic syndrome.

Immunosuppressive treatment after successful renal transplantation (age 6) was with azathioprine and prednisolone. The primary renal disease (congenital nephrotic syndrome) was considered to be an unlikely pretransplantation cause of measles immunisation. At 10 years of age the child had a ‘flu-like illness with morbilliform rash. Three months later she suffered a second ‘flu-like illness and within days, developed myoclonic seizures involving arms, head, and subsequently legs. An electroencephalogram showed periodic and stereotyped bursts of high voltage slow components accompanying the clinical events. Seizures were absent during sleep; consciousness and intellect were preserved when awake. Anticonvulsant treatment was ineffective. Intraethelial synthesis of measles virus antibody was documented (serum titre 40, partial viral fluid 4 in presence of normal blood cerebrospinal fluid barrier function; cerebrospinal fluid/serum albumin 2 4+10-4). Rubella virus antibody (serum 640, cerebrospinal fluid <1, serum of rubella virus IgM only weakly positive) was also found at this time but neither antibody was detectable in serial serum samples collected prior to the first ‘flu-like illness. Antibody titres were unchanged in a subsequent serum sample suggesting both measles and rubella to be temporarily associated with the first illness (the second remained undiagnosed). Four months after the onset of seizures she developed chickenpox, lapsed into coma, and died six weeks later without regaining consciousness. Chickenpox probably accelerated the clinical course but not by direct central nervous system invasion (serum Herpesvirus varicellae titre 200, cerebrospinal fluid <1).

It is possible that rubella either triggered a latent measles virus infection (although we found no serological evidence of previous measles) or, interfered with the normal measles immune response (compounding the effects of immunosuppression) leading to progressive central nervous system infection.³ Whatever the mechanism, this case reinforces the view that all children given therapy for renal transplantation should receive measles immunisation.

P E KLAPPER
G M CLEATOR
Division of Virology, Department of Pathology, Manchester Royal Infirmary, University of Manchester

M C CLARKE
J POSTLETHWAITE
Departments of Pathology and Renal Medicine, Booth Hall Children's Hospital, Manchester, United Kingdom


Refux vomiting

Sir,—It was kind of Dr Mills to refer to some of my publications in his article on reflux vomiting.¹ Unfortunately some of his interpretative conclusions require correction. Like many other writers on this subject he has totally misrepresented my 1959 observations on the natural history of vomiting infants with a partial thoracic stomach (hiatal hernia) as being applicable to all infants with ‘symptomatic gastro-oesophageal reflux’. This is most certainly not so. His quote relating to the incidence of gastro-oesophageal reflux is equally misleading as the hospital estimate of one in 500 to which he refers relates to the hospital attendance of children with a partial thoracic stomach and not to that of children with reflux per se as mentioned in his article. Clear separation of the natural history of reflux with its sequelae and reflux without these needs to be made.

Refux in infants with a partial thoracic stomach may be accentuated by a host of factors including stress, feeding volume, and gastric dysfunction. Liquid volumes less than 100 mlare unlikely to cause reflux and its presence in infants with a small gastric capacity is as misleading as the statement that ‘The finding of a hiatus hernia with or without an associated partial thoracic stomach is not of itself an indication for surgery’.¹ I have tried substituting gastro-oesophageal reflux for hiatus hernia as a useful clinical indicator of a partial thoracic stomach but in neither instance does this alter the meaning of the phrase in the particular context of his article.

Largely as a result of the emphasis placed on the importance of gastro-oesophageal reflux per se and its detection by non-radiological means very much less attention is directed nowadays to the clinical picture of infantile reflux with a partial thoracic stomach, which is often regarded as having little clinical relevance. I have, however, found on long term clinical evaluation of many hundreds of infants with reflux that the presence or absence of a partial thoracic stomach serves as a very valuable clinical guide to prognosis.² For whereas reflux in infants with a partial thoracic stomach is
associated with a significant morbidity, gastro-
oesophageal reflux in the absence of this anom-
aly is a relatively benign self-limiting condi-
tion, which in the case of vomiting infants can
usually be treated successfully by appropriate
and adequate thickening of feeds. I am there-
fore in no doubt as to the clinical importance
of distinguishing infants with reflux and a
thoracic stomach (in whom the antireflux
contribution of the abdominal oesophagus is
absent) from those with reflux as the only
observed abnormality. Such reliance on the
prognostic significance of a partial thoracic
stomach is of course totally dependent on hav-
ing an experienced paediatric radiologist as a
colleague who is equally aware of the import-
ance of carefully examining infants with reflux
for a partial thoracic stomach.

1 J Carré, Le Vat é, Los Bliqü, St Andre's, Gourou,
Channel Islands

Growth after gut resection for Crohn's
disease

Sir,—We read with interest the paper by McLain et al, and we would like to comment on both the methodology of their data collec-
tion and the authors' conclusions. Firstly, this was a retrospective study, and thus has the major disadvantage of lacking standardisa-
tion in the collection of the auxological data.
There is no mention in their paper of how height measurements were recorded and stan-
dardised, or on the accuracy of the staging of pubertal development.

Secondly, while agreeing with their findings that a dramatic growth acceleration may occur after surgery for Crohn's disease, we disagree with their conclusion that 'catch up growth is not limited by the stage of puberty'. We have recently completed a large, prospective study involving the factors influencing growth after bowel resection for Crohn's disease in 42 chil-
dren requiring surgery before their 17th birth-
day. All growth indices (including height meas-
urements every three months) were per-
formed prospectively by a clinical auxologist,
and an accurate pubertal staging was per-
formed in each patient at the time of surgery,
together with the radiological bone age in the
majority of cases. Our data (table 1) clearly
show a strong relationship between height
velocity in the first postoperative year and the
pubertal status (Tanner breast and genital
d stage) at time of operation. The apparent
lack of effect of pubertal status on growth in the
study of McLain et al may merely reflect the
even small number of children in advanced pubertal
stage (one), or the retrospective nature of their
data collection. In our opinion, the tim-
ing of surgical intervention is vital, and should
be performed before puberty becomes too
advanced and the potential for catch up
growth is lost. This is in direct contrast to the
conclusion stated in their paper.

Finally, we monitor growth very carefully
in all our children with Crohn's disease. Any

<table>
<thead>
<tr>
<th>Boys</th>
<th>Girls</th>
</tr>
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<tbody>
<tr>
<td>Before surgery</td>
<td>After surgery</td>
</tr>
<tr>
<td>Prepubertal: stage 1 (n=18)</td>
<td>1.95</td>
</tr>
<tr>
<td>Early puberty: stages 2 or 3 (n=14)</td>
<td>2.81</td>
</tr>
<tr>
<td>Late puberty: stages 4 or 5 (n=10)</td>
<td>3.32</td>
</tr>
</tbody>
</table>

*p<0.001, t̅p=not significant.

Mean preoperative and postoperative height velocities (cm/year) in 42 children undergoing bowel resection for Crohn's disease grouped according to puberal status at time of operation


Drs Davidson and Beasley comment: Thank you for the opportunity to reply to the correspondence of Evans et al. We agree com-
pletely with their comments regarding the relationship of the timing of surgery to puberty on catch up growth in Crohn's dis-
ease. Although we acknowledge that we have insufficient patients in advanced puberty to comment on the effect on growth of the stage of puberty, our data would suggest that catch up growth is not limited by the onset (as dis-
tinct from stage) of puberty. The one patient in advanced puberty clinically who exhibited catch up growth after surgery did not have his bone age assessed.

Like Evans et al we would emphasise the importance of careful ongoing growth mon-

Finally, we are pleased to find that the pros-
pective data of Evans et al support our ret-
rospective observation.

S P R I N G  B O O K S

The paediatric departmental library

What should be the priority for the paediatrician when financial considerations reign and access to current literature is still a battle for books and journals? The rate of change in paediatric practice and slowness of publication in jour-

The paediatric departmental library

What should be the priority for the paediatrician when financial considerations reign and access to current literature is still a battle for books and journals? The rate of change in paediatric practice and slowness of publication in jour-
nals and especially in textbooks lead to such a rapid obsolescence that departmental libraries are rarely of use for any in depth study. They provide a source to back up basic case presen-
tations but at a considerable expense. It is impossible to keep up to date with the rapidly changing field of knowledge. There is no other way to incorporate a totally up to date reference in your daily practice and still at a reasonable cost.

There must be the equivalent of a paediatric encyclopedia covering all major subject
areas, such as the Textbook of Paediatrics by Forfar and Arneil or Nelson's Textbook of Paediatrics. Then a selection of system or dis-

Better way of updating, but still relying on books and journals is the use of a file of reviews articles from paediatric journals or free journals such as Hospital Medicine, Medicine International, Update, Hospital Update, Prescriber's Journal, and Drugs and Therapeutics Bulletin. The difficulty is the archiving and security. However the avail-

Availability of photocopiers has made this easier, although care must be taken to avoid infringe-
ing copyright laws. Another very useful practice is to incorporate a copy of a key article in the hospital notes of the patient with this particular diagnosis. It is important to keep updating this and to include this article in the departmental "useful literature" file. The filing system is unlikely to succeed unless it is clearly on individual's responsibility and that should be at fairly senior level.

However, we are on the brink of the break-
through in data retrieval which has had the Medline searches and now the CD ROMs as forerunners. When the computer terminal is as familiar and essential an item on the desk in consulting room and ward rooms, you will be able to access up to date original articles and learned reviews at the touch of a few buttons. I can foresee the time when the medical details (plus accounting data) is entered into the consulting room computer an automatic search will be made for relevant new literature on that child's condition. A summary of this will be available for inclusion in the word pro-
cessed report that will be available for any of the child's professionals to access, as well as for inclusion in the hardcopy that the parents will be carrying. So the medical library will pass into the mists of memory as the stock mar-
kets runners have and as the filing rooms in hospital will.

GR AHAM C LAYDEN
Reader in paediatrics


Who needs a Filofax when he can have the vade-mecum? This remarkable little book must have a greater concentration of paediat-

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