LETTERS TO THE EDITOR

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 unoxygenated 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). 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, the authors state that five of the eight had concurrent pathological processes that are independently associated with sudden death, and in some cases they had cyanosis: intraesophageal bolus of food 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). It would strengthen the underlying hypothesis considerably if further information could be provided detailing how these well-established causes of sudden death were eschewed in favour of intrapulmonary shunting as the primary 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 the issue 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


Des 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 intraesophageal impacted food bolus, and a brainstem glioma were 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 hypoxic episodes having charactristics 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 remains 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, no 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. The wisdom of current recommendations1 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 a late complication of pretransplant 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 in 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. Intrathelial synthesis of measles virus antibody was documented (serum titre 40, partial thoracic stomach serves as a very valuable clinical guide to prognosis.2 For measles reflex vomiting in infants with a partial thoracic stomach is


Reflex vomiting

Sir,—It was kind of Dr Mills to refer to some of my publications in his article on reflex 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. Clearly the number of infants with a partial stomach with reflux and no recognisable displacement into the thorax of the gastro-oesophageal sphincter would be greatly in excess of this figure. I am also perplexed by his statement that ‘The finding of a hiatus hernia with or without an associated partial thoracic stomach is not of itself an indication for surgery’. I have tried substituting gastro-oesophageal reflux for hiatus hernia and partial thoracic stomach but in neither instance does this alteration clarify the meaning of this sentence 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 ‘flu-like illness. Anti-body 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 antibody titre 1:320, 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, in the absence of 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 go on to renal transplantation should receive measles immunisation.

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

M C CLARKE
R J POSTLETHWAITE
Department of Neurology
and Renal Medicine,
Booth Hall Children’s Hospital,
Charlesworth Road,
Blackley,
Manchester M9 2AA

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R W Byard

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