may deserve further study.\textsuperscript{8}\textsuperscript{15}\textsuperscript{16} provided investigators—even in the face of daily and severe attacks—can withstand the temptation to ‘resuscitate’ or give uncontrolled treatments.

Insufficient attention has been paid to the neurological basis of the active expiration which is an essential feature of breath holding attacks, but which may also be seen in those pallid syndromes (associated with cardiac asystole) induced by ocular compression in the laboratory.\textsuperscript{13} Obviously the oculorespiratory reflex is no more involved in normal breath holding than is the oculocardiac reflex in those ‘reflex anoxic seizures’ mediated by cardiac asystole, but it can be studied safely and ethically\textsuperscript{17} and it is not confined to man. What is self evident is that if continued brainstem neuronal drive is necessary to inhibit active inspiration and/or cardiac systole, then sufficient hypoxia and/or ischaemia must reverse this inhibition.

Summary

In their recent publication in this journal, Southall et al described typical cyanotic breath holding spells, both in otherwise healthy children and in those with brainstem lesions and other malformations.\textsuperscript{1} Their suggestions regarding possible autonomic disturbances may require further study, but they have adduced no scientific evidence to contradict the accepted view that in the intact child blue breath holding spells are benign.

Those families in which an infant suffers an ‘apparently life threatening event’ deserve immense understanding and help, and it behoves investigators to exercise extreme care and self criticism in the presentation of new knowledge which may bear upon their management and their morale.


Cyanotic ‘breath holding’ and sudden death

M P Samuels, D G Talbert, D P Southall

Dr Stephenson raises an important issue concerning the risk of sudden death during cyanotic breath holding. Our recent paper describes investigations to understand further the pathophysiological mechanisms responsible for severe cyanotic breath holding and did not attempt to estimate the risks of this disorder.\textsuperscript{1} To do this would require a population based study, ideally prospective in nature, in which there was almost complete follow up. The historical studies mentioned by Dr Stephenson are unable to confirm that no deaths occurred. Thus in the study of Lastal et al\textsuperscript{2} and Lombroso and Lerman,\textsuperscript{3} 18% and 43% respectively were lost to follow up. One possible reason for a lack of response to posted questionnaires might have been the death of a child.

We trusted that readers would not automatically infer from reading our paper that there is mortality of 16% in cyanotic breath holding. As stated, our patients were at the severe end of the spectrum and therefore not representative of the whole population of ‘breath holders’. Furthermore, Hunt in his recent editorial did not claim that 15% of breath holders die, but was specifically referring to our collection of patients.\textsuperscript{4} Our cases were ‘selected’ by virtue of their referral to us by their paediatrician who, with the parents, was concerned about the severity of the hypoxaemic episodes.

Dr Stephenson omits to mention our own prospective population based study of sudden deaths between 1 and 5 years of age.\textsuperscript{5} This involved the follow up of 9856 infants all of whom were tagged for future death by the Office of Population Censuses and Surveys. Two of the deaths between 1 and 5 years in these 9856 children occurred during cyanotic breath holding. Based on Lombroso and Lerman’s estimated incidence of cyanotic breath holding as 2–8% of all children,\textsuperscript{2} around 276 of our 9856 cohort should have suffered from this
disorder. Thus around two of 276 or 0.7% of children with cyanotic breath holding are likely to have a fatal outcome. If one can accept a 0.7% mortality rate, statements concerning the benign nature of this condition are correct.

There are three other reasons why we consider that the severe forms of cyanotic breath holding are not benign. Firstly unconsciousness is due to severe cerebral hypoxia and this highlights the danger of adopting a conservative approach to the management of patients at the worst end of the spectrum of this disorder.

However, asystole. reported profound respiratory reflexes.6 forces and reference are now of noradrenaline an effective way to avoid endotracheal intubation. Dr sure, Dr Stephenson's advice to be inappropriate; eight of 51 such cases reported in our paper died. Since our publication we have experienced an artifical hypoxia and this highlights the danger of attempting resuscitation. For the majority of young children with isolated and infrequent episodes we would agree with this opinion. However, for those with frequent episodes involving loss of consciousness, we consider Dr Stephenson's advice to be inappropriate; eight of 51 such cases reported in our paper died. Since our publication we have experienced an artifical hypoxia and this highlights the danger of adopting a conservative approach to the management of patients at the worst end of the spectrum of this disorder.

This infant presented at 6 weeks of age with severe hypoxaemic episodes which were clinically recognised as cyanotic breath holding. Investigations in our department confirmed the presence of active respiratory 'apnoea' and intrapulmonary shunting. His parents were given a transcutaneous PO2 monitor to identify any episodes that should occur when he was not being observed. His episodes had always occurred when he was awake and his mother did not believe that they would occur when he was sleeping. Nevertheless the episodes were considered by us and his referring paediatrician to be extremely dangerous and it was explained to his mother that an episode might occur if he awoke from sleep and became frightened or angry. One afternoon at the age of 10 months, he was settled to sleep in his cot (unattached to the oxygen monitor). Forty five minutes later his mother found him dead. A postmortem examination showed patchy congestion in the lungs with some milk in the larger airways; these findings were reported to be compatible with sudden infant death syndrome, although, because of his previous history, his death was certified as due to 'apnoea of infancy'.

In summary, the episodes of hypoxaemia described in our paper represented the severest form of cyanotic breath holding (a term which is unfortunate in that it implies a voluntary component). A functional disturbance, which involves intrapulmonary shunting and prolonged respiratory apnoea, represents the best explanation for the final pathophysiology leading to the sudden and unexpected deaths that occurred in eight of our 51 patients, despite the pathology found in some at postmortem examination.7 The risk of sudden death in all patients with clinically recognised cyanotic breath holding appears to be very low, but we would continue to be concerned by episodes in infants less than 6 months of age or in those with frequent loss of consciousness. The identification of an effective treatment will not only help in our understanding of the mechanism responsible for these episodes, but may also improve the lives of many young infants and their families.

Cyanotic 'breath holding' and sudden death.

M P Samuels, D G Talbert and D P Southall

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