Pallid syncope (reflex anoxic seizures)

Breath holding attacks are familiar to family doctors and paediatricians as non-epileptic episodes occurring in some infants and young children as a consequence of a physical or psychological hurt. In the common cyanotic type, diagnosis is not difficult if an adequate history is available, although the occasional occurrence of clonic movements and incontinence may raise doubts that the attack is primarily epileptic. (Such doubts may be dispelled if the time relation to the hurt is clear—reflex epilepsy is very rare except in the light-sensitive form.) More uncertainty is present if breath holding is merely a gasp with no preceding crying, if there is severe opisthotonos, or if the child is limp and pale. In these circumstances again it is the history of the preceding event that is diagnostic.

Distinction between cyanotic and pallid breath holding

An important study of breath holding 'spells' was published 17 years ago by Lombroso and Lerman.¹ They made a clear distinction between the cyanotic and the pallid forms, both on history and on the evidence from their own and previous studies that a different mechanism was involved in the two types. Whereas the cyanotic type of breath holding attack is due to cerebral ischaemia from a sudden rise in intrathoracic pressure impeding venous return to the right side of the heart (the Valsalva manoeuvre), the pallid type is due to vagal asystole. They showed, following the work of Gastaut and Gastaut² and others in the previous decade, that vagal stimulation by eyeball compression was much more likely to produce prolonged asystole in children with pallid attacks than in those with cyanotic ones. Stephenson³ has emphasised the usefulness of this manoeuvre, both diagnostically and therapeutically (by demonstrating the mechanism of the attack to the parent).⁴ He has also shown it to be safe. Asystole, even up to 32 seconds in duration, has produced no evident complications and he has never had to use resuscitatory methods.

Making the diagnosis

Most paediatricians will nevertheless be reluctant to use the oculocardiac reflex, certainly without electroencephalographic and electrocardiographic monitoring, even if reassured of its safety. Eyeball compression may seem repugnant, even punitive,

and damage to the eyes, perhaps in the long term, cannot be excluded. Lombroso and Lerman¹ do not recommend the manoeuvre in cases of high myopia. How often is it necessary? In my experience a careful description of the episode and its antecedent circumstances is usually sufficient for diagnosis. The pallid attack often occurs in infancy and follows a few seconds after a sudden, unexpected, mildly unpleasant stimulus. A moderate blow on the occiput (that is, without visual warning), being placed in the adult bath for the first time, and the introduction of a new taste⁵ are examples of such stimuli and contrast with the more violent and painful stimuli liable to produce a cyanotic breath holding attack. While the pallid attack is understandable as a vagally induced 'escape' mechanism (as in the syncope of older children and adults) the cyanotic attack seems to be a rage reaction after a bellow of pain or frustration. The pallid attack also resembles the more familiar syncope in the observed features: the extreme pallor, the 'cold sweat', the total limpness, and the appearance of death followed by a fairly rapid return to normality. Opisthotonos, a few clonic twitches, and turning up of the eyes are fairly common, however, and may raise doubts—almost always unjustified. If the clinical description is atypical or incomplete it may be helpful to defer diagnosis until a more careful observation has been made and the description put in writing immediately afterwards. It is rare in my experience for the diagnosis to remain obscure after one or more careful histories have been taken, but occasionally a conventional electroencephalographic examination is justified.

Diagnosis is very important, mainly to remove the label of epilepsy or convulsions and in some instances the anticonvulsant drug already being given. Many family doctors are unaware of the existence of infantile pallid syncope and, therefore, find it difficult to disregard the possibility of epilepsy in an infant whose episodes do not resemble the common cyanotic breath holding attacks.

Terminology

The matter is made more difficult by the terminology. In the United States, and increasingly in Britain, the term 'seizures' is used to describe epileptic phenomena. I regard the term 'reflex anoxic seizures' as a most unfortunate one and much prefer 'pallid syncope' which is descriptively and

physiologically correct as well as being more informative and reassuring to parents. An acceptable alternative, although less informative to them, is 'vagal attack' as suggested by Stephenson³ but he does not use it in his latest publication.⁶ Movements, particularly opisthotonic, there may be, but these are not the result of massive synchronous bio-electrical cerebral discharges—that is epilepsy.

Use of atropine

The most important treatment is to explain to the parents what the episode is, what it is not, and the prognosis—which is excellent. Occasionally, however, a course of atropine has also been found to be necessary. Lombroso and Lerman mentioned it in 1967¹ without giving details and recently McWilliam and Stephenson⁶ described its use in seven children with unusually severe or frequent episodes. The latter's results were impressive in reducing the number of attacks, with none of the side effects of atropine. This drug (atropine methonitrate was preferred to atropine sulphate) is, therefore, a useful treatment for the occasional patient and might occasionally give supportive evidence of the diagnosis, but overdosage may occur if the details of administration are not understood.

Frequency

How common is it? In a follow up study of nearly 5000 deliveries at the Boston Lying-In Hospital, Lombroso and Lerman found the incidence of breath holding attacks to be 4.6% and one fifth of

these (1%) were pallid syncope. This suggests that the condition is more common than most paediatricians think. If the suggestive evidence that some 'febrile' convulsions are in reality pallid syncope⁷ is confirmed, then it must be even more common.

It is, therefore, important for paediatricians to make the condition more widely known to family doctors, clinical medical officers, and others concerned with the care of preschool children. An important, helpful step which could be taken now would be to agree on 'pallid syncope' as the preferred term for this condition and abandon the misleading 'reflex anoxic seizure'.

References

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