Reflex anoxic seizures

May be frightening to parents but are nothing to worry about

A reflex anoxic seizure is a paroxysmal event triggered by a painful or frightening stimulus which, by vagal stimulation, causes pronounced bradycardia or cardiac arrest and consequent relative cerebral ischaemia, thereby inducing an anoxic "seizure" or "attack".1 2

A typical attack is always provoked, although the provocation may not be witnessed. The most common trigger is sudden pain or fright, such as that due to trapping a finger in a door or a minor blow to the head, particularly the occiput; other provoking factors include febrile illnesses,4 venepuncture,5 and an excessively hot or cold bath. The painful experience may be accompanied by a gasp or brief cry, after which the child falls, limply, and appears pale, even deathly white, "as though dead." This phase corresponds to the period of asystole, which is brief, usually lasting less than 15 seconds. The child may then rapidly return to normal with or without crying, although she may sleep.

Frequently, though not invariably, stiffening of the body or opisthotonus, clonic movements, upward deviation of the eyes, and urinary incontinence may follow the initial limp phase.6 It is these features that may lead to a misdiagnosis of epilepsy, particularly if the painful or frightening trigger is not witnessed. Stephenson believes that the anoxic seizure may, rarely, induce an epileptic seizure,7 although I and other authors have never known this to happen.

Attacks may occur very infrequently or many times a day; this may reflect the extent of the vagal hyperexcitability or the degree of the noxious stimulus, or both. Infants and young children (aged 6 months to 2 years)7 are particularly susceptible, but the condition may occur in adolescents and even adults, and in other family members.8 Girls are more commonly affected. The condition is common and probably underdiagnosed; epilepsy and (blue) breath holding attacks are the most common misdiagnoses. Occasionally, sudden pain may induce tonic-clonic seizures in patients with epilepsy, but this should be easy to recognise. The precise number of children who experience these attacks is unknown, given the degree of misdiagnosis (one study has estimated that about 8/1000 preschool children are affected).8 The condition may be as common as epilepsy.

The diagnosis is made on the basis of an accurate history (which may be difficult to obtain if the onset was not witnessed). Even when the entire attack has been seen, witnesses frequently recall only the stiffness or abnormal movements, leading to a misdiagnosis of a convulsion or grand mal fit and prescription of antiepileptic drugs.9 If the history is incomplete or atypical a written description of further events should be sought from parents or other witnesses to establish the diagnosis. Ocular compression has been suggested as a useful diagnostic and therapeutic manoeuvre in those children in whom the condition is suspected or if families require further reassurance.4 Ocular compression stimulates the vagus nerve (the oculocardiac reflex), leading to bradycardia, asystole, and, in the electroencephalogram, high voltage slow activity with or without a period of flattening and a clinical attack (the so-called anoxic seizure, or syncopal attack).1 2

Ocular compression may be distressing for the child and must be undertaken with simultaneous electrocardiographic and electroencephalographic monitoring for obvious reasons. The technique should be avoided in children with a known cardiac arrhythmia or glaucoma and in units with limited experience of it. Some authors consider that ocular compression is unnecessary;1 6 they believe that the condition can be diagnosed on the basis of the clinical description, and parental reassurance does not depend on a demonstration of the phenomenon, even in a controlled situation.

The phenomenon is benign; the period of cardiac asystole is brief and children do not die or suffer cardiac or cerebral damage. Limited evidence suggests that resolution of the
attacks usually occurs by early childhood; occasionally the episodes may persist into early adult life.

The most important aspect of treatment is to explain the condition to parents and to reassure them that it is not epilepsy and that the prognosis is good. Atropine has been advocated to prevent the vagal hypersensitivity in those children with frequent, persistent attacks or when parents remain concerned. Although atropine may be effective, its pharmacodynamic properties indicate that dosing may need to be frequent, which increases the risk of overdose. Drug treatment has been unnecessary in my and at least one other author's experience. 

"Reflex anoxic seizure" is not universally accepted as the most appropriate term for this phenomenon because the word seizure in many medical (and non-medical) minds is synonymous with epilepsy. As the condition is often misdiagnosed as epilepsy (this accounted for 15% of misdiagnoses in one series) the description of the attacks as seizures should perhaps be avoided to prevent any additional confusion or concern. Parents often remember only the word seizure and not what it means, no matter how careful the explanation. Because of this, and the suggested mechanism, these attacks have also been called white or type 2 breath holding attacks (in contrast to the blue or type 1 breath holding attacks), although the primary event is not one of breath holding. Other names include reflex anoxic cerebral seizures with white syncope, which is too verbose a label, and pallid infantile syncope or pallid syncopal attacks, which is probably the most popular alternative.

An awareness of the condition and an accurate history are crucial to both diagnosis and "treatment." This may help to resolve some of the concerns that has been expressed recently in the national press and that has prompted the inception of a self help group for "afflicted" families.

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Who needs dual chamber pacing?

The British guidelines probably exaggerate the needs; proper studies should be done

Traditional cardiac pacing transformed the lives of a generation of patients with heart block. Victims of Stokes Adams attacks regained their confidence and their driving licences. Those with fatigue and heart failure due to bradycardia were improved. Their pacing systems were simple: a single transvenous electrode positioned in the right ventricle, which was attached to a generator implanted subcutaneously. It paced the heart at 70 beats per minute and was inhibited if the patient's own heart rate exceeded that speed.

These patients fared well. But experience over the past 15 years suggests that they might have fared even better with a more complex system. The addition of an atrial electrode to sense or pace the atria (or do both), linked to a dual chamber pacemaker that then triggers ventricular depolarisation, has many theoretical and practical advantages. Firstly, restoring atrial transport improves stroke volume; secondly, heart rate can respond to effort and further improve exercise capacity provided that sinus node function is normal; thirdly, compared with single chamber systems it may reduce the risk of the subsequent development of atrial fibrillation and systemic embolism; fourthly, the pacemaker syndrome—a vague collection of symptoms due to the atria and ventricles contracting asynchronously—is abolished; and finally, dual chamber pacing is now reliable and effective over years.

The only definite contraindication to dual chamber pacing is established atrial fibrillation, but even then a heart rate response to exercise can be achieved in those with a bradycardia by implanting a generator that senses a bodily change—for example, movement—and accelerates accordingly. Dual chamber and rate response devices are but two of the more spectacular developments in pacing technology. These and others, such as the ability to reprogram the generator after implantation, have led to such an array of alternatives that in 1991 two groups of experts published guidelines for pacemaker implants, taking into account both the technical advances and the widening indications for this form of treatment. Both the British and North American reports recommended the greater use of more complex pacemakers; the document from the British Pacing and Electrophysiology Group contains the suggestion that "the atrium should be paced/sensed unless contra-indicated." This has financial consequences, as the authors predicted.

A dual chamber pacing system costs almost three times as much as a simple ventricular system—about £2100 compared with £750 (1991 prices). Wholesale implementation of the British recommendations would increase pacing budgets by an estimated 75% to 94% although this has been challenged. The expense does not stop there, however; dual chamber systems are more difficult to insert; follow up appointments take longer; and pacemaker malfunction is more difficult to detect, placing greater demands on technical staff. In addition, the generators will probably not last as long.

The working party of the British Pacing and Electrophysiology Group and others have argued that costs should not inhibit the promulgation of an ideal pacing practice. Quite so, but a more serious objection is that all the guidelines ignore patients and their clinical needs; the recommendations deal solely with conduction systems and their defects.

The average age of the 700 patients who received new pacemaker implants at this hospital in 1990-1 was 76 years; 82% of the patients from St George's Hospital, London, were over 65. Most pacemaker patients are elderly. Their exercise