Syndrome X and hyperventilation

Sir,—Syndrome X and hyperventilation have long been associated, and Lewis and colleagues (British Heart Journal 1991; 165:94—6) are to be congratulated for their discriminating contribution to the field and for raising several important questions. A central issue is whether hyperventilation can be dismissed on the basis of a rather limited schedule of testing. For example, it is no longer thought that the forced hyperventilation provocation test (FHPPT) is the "gold standard" of hyperventilation testing,1 but that provocation by personally relevant stressors2 is at least as important. Indeed, in the context of cardiac patients, the latter stressors, which can be used to activate the sympathetic nervous system (with consequences such as vasocostricture pathways)3 are probably the triggers most relevant in everyday life. Furthermore, besides the interplay between hyperventilation and sympathetic nervous system activation (with consequences such as"magnesium depletion") it seems likely that it is the neuroendocrine setting that can determine whether or not a given episode of hyperventilation has vasocostrictive consequences.

It is a commonplace finding that many patients with recurrent hyperventilation illnesses do not hyperventilate during an exercise test, but readily overbreathe in response to an emotional challenge, particularly when the challenge involves the recall of feelings being trapped or of anger.4 As far as the data presented by Lewis et al are concerned, besides noting the absence of figures for end tidal pressure of carbon dioxide (PETC02) for their controls, we also note that the PETC02 values of 38 mm Hg at 50% of maximum exercise and 37 mm Hg at maximum exercise are well below expected normal values.5 The demonstration of increased minute ventilation for given minute carbon dioxide (VE/VC02) is quite consistent with chronic hyperventilation. It is due either to reduced respiratory centre buffering6 or a flywheel effect which regularly over-activates pathways promote breathing,7 or both. The observation of a raised VE/VC02 may well be the key indicator of a longer term tendency to hyperventilation, because as stated above, the FHPPT is a single act of hyperventilation and is dependent upon the patient's starting point in terms of other influences upon vasoconstriction and upon the body's buffering systems.

A basic question about syndrome X is whether their responsiveness to recognised vasocostrictive influences is greater than average, just as Europeans in general are more responsive than the Japanese.11 We suggest that Lewis et al have sharpened the definition of characters in the drama of syndrome X and hyperventilation but have not yet brought down the final curtain.

STUART D ROSEN J C KING P G F NIXON
Cardiac Department, Charing Cross Hospital, Fulham Palace Road, London W6 8RF

3 Lewis BI. Chronic hyperventilation syndrome. JAMA 1964;189:404-8.

This letter was shown to the authors, who reply as follows:

Sir,—We appreciate the interest shown by Dr Nixon and colleagues in our paper. Our study refers of course to carefully characterised patients—an important consideration given the widely differing patient groups represented in published reports on patients with unexplained chest pain. Clearly the filing category of syndrome X is coming to the end of its useful life, particularly in view of the confusion now arising over a different "syndrome X".

Sir,—We make no comment on the points raised by Dr Nixon and colleagues—our patients with syndrome X all described typical angina and, in every case, exercise induced the same symptom. We would argue that an exercise test in such a group represents a "personally relevant stressor". The finding of normal arterial Pco2 values throughout exercise in our syndrome X patients formed the basis for our conclusion that chronic hyperventilation was not inappropriate (which is in common usage is the implication inherent in the term). Because we showed that end tidal Pco2 correlated only poorly with arterial Pco2, in these patients, their end tidal measurements, we agree, provided no evidence either for or against a diagnosis of hyperventilation. Chronic hyperventilation may indeed increase the VE/VC02 slope but only in the presence of low arterial Pco2 (the modified alveolar gas equation states: VE = Vco2/Paco2-1 (Vd/Vt)). The normal arterial Pco2 in our patients suggests an increase in deadspace ventilation.2 We did not measure arterial Pco2 in our control patients for ethical reasons.

We share the view that the final curtai n is not yet drawn on the various players on this ill-lit stage, while our spotlight continue to weaver....
Echocardiographic diagnosis of a ruptured aneurysm of the sinus of Valsalva: operation without catheterisation in seven patients

Stir,—Dr Sahasakul and colleagues (British Heart Journal 1990;64:195–8) clearly described the echocardiographic diagnosis of a ruptured aneurysm of the sinus of Valsalva, but they made no suggestion about its aetiology or any thoughts to label this a disease of "congenital origin". The ruptured sinus affected the right coronary sinus in six of their seven patients. Figure 1 in their article clearly shows the absence of the subpulmonary infundibulum and fibrous continuity between the aortic root and the pulmonary valve (fig 1B), features characteristic of a doubly committed subarterial ventricular septal defect, with protrusion of the right coronary sinus into the right ventricle (fig 1A).

The usually accepted explanation for a congenital aneurysm of the sinus of Valsalva is thinning of the wall of the aortic sinus just above the leaflet hinge, although this alone does not explain why the left coronary sinus is hardly ever involved. The association of the aneurysm of the sinus of Valsalva with a ventricular septal defect was described in Japan. Several changes in morphology which were proposed, leading to rupture of the right coronary sinus into the right ventricle in young adulthood when there was a ventricular hinge in the outlet portion of the septum.1 It is notable that there is an increased incidence of ruptured aneurysm of the sinus of Valsalva in Oriental populations, just as there is of doubly committed subarterial ventricular septal defect. Large defects without deformity or offsetting of the aortic valve cause major haemodynamic disturbance in infancy, whereas smaller holes typically present in childhood with aortic regurgitation because of prolapse of the right coronary aortic cusp; but they may present with rupture of a sinus of Valsalva aneurysm in adult life.4 Indeed, during a 10 year period at the Japanese Red Cross Hospital 32 of 36 patients undergoing surgery for a ruptured aneurysm of the sinus of Valsalva had "subarterial infundibular" ventricular septal defects.5

Thus it should be emphasised that the likely substrate for aneurysm of the sinus of Valsalva in many patients is the presence of a doubly committed subarterial ventricular septal defect with offsetting of the arterial valves. Prolapse of the right coronary aortic cusp results in elongation of the sinus and thinning of the wall just above the leaflet hinge. Mechanical stress leads to dilatation of the sinus over many years, and sometimes rupture into the right heart. Patients with a doubly committed subarterial ventricular septal defect and offsetting of the arterial valves, which can be observed echocardiographically, are at risk of this later complication.6

IAN D SULLIVAN
MARC DE LEVAL
Hospital for Sick Children, Great Ormond Street, London WC1N 3JH


This letter was shown to the authors, who reply as follows:

Stir,—Dr Sullivan and Dr de Leval have correctly pointed out the association between a ruptured aneurysm of the sinus of Valsalva and a doubly committed subarterial ventricular septal defect in most cases reported in the Orient. Nevertheless, there are several other causes of a ruptured aneurysm of the sinus of Valsalva, chiefly a congenital lack of fusion between the media of the aorta and the annulus of the aortic valve.7 Other reported causes include bacterial endocarditis, Marfan's syndrome, coarctation of the aorta, bicuspid aortic valve, trauma, and also the association with ventricular septal defect.5,8

The reason for the high prevalence of ruptured aneurysms of the sinus of Valsalva in Oriental patients is not known. We propose a genetic cause or perhaps a logistic one: patients with ventricular septal defect in the Orient may not see a doctor until symptoms develop—that is after the aneurysm has already ruptured. The latter proposal is particularly true in Thailand because many paediatric patients with ventricular septal defect went unnoticed for several years because they were born in remote provincial areas without modern medical facilities.

YONGYUTH SAHASAKUL
SUPHACHAI CHAITHIRAPAN
Division of Cardiology, Siriraj Hospital, Mahidol University, Bangkok 10700, Thailand


Impending paradoxical embolism: a rare but important diagnosis

Stir,—I read with interest the recent case report of impending paradoxical embolism by Speechly-Dick et al. (British Heart Journal 1991;65:163–5). However, I would like to offer two differing opinions on the points they made.

First, their statement that only two of 37 reported cases of paradoxical embolism were diagnosed during life was an underestimation. All of 25 cases reviewed by Leonard et al10 and 27 cases reviewed by Jungbluth et al, cited by the Speechly-Dick et al in their paper as references 4 and 6, respectively, were diagnosed during life. The two cases reported by me in 1976 were diagnosed during life.2

Second, the current treatment of choice for patent foramen ovale or minute atrial septal defect as a cause of paradoxical embolism is non-surgical transcatheter closure.3 If this defect could be closed during a cardiac catheterisation, the need for open heart surgery would be obviated together with the extended hospital stay, expense, need for reoperation, and morbidity, mortality, and psychological trauma associated with surgery.

TSUNG O CHENG
Division of Cardiology, Department of Medicine, The George Washington University Medical Center, Washington, DC 20037, USA


This letter was shown to the author, who replies as follows:

Stir,—Professor Cheng is correct in saying that there are many cases of paradoxical embolism diagnosed in life and reported in the literature. However, our case report described the rare finding of impending embolism of which, to my knowledge, there are only two previously reported cases. The second point which Professor Cheng raised was that in his opinion the current treatment of choice for patent foramen ovale or minute atrial septal defect is non-surgical transcatheter closure. This is a relatively new procedure for which we have, as yet, no long term results. Surgical closure, however, is a well-established and accepted technique with very low mortality and morbidity and excellent long-term results.1 Non-surgical transcatheter closure may well be the treatment of the future but is not yet the treatment of choice.

M E SPEELEY-DICK
Department of Cardiology, Middlesex Hospital, London W1N 8AA