Syndrome X and hyperventilation

Syndrome X—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 (FHT) is the "gold standard" of hyperventilation testing, but that provocation by personally relevant stressors is at least as important. Indeed, in the context of cardiological patients, the latter stressors, which can be known to activate the energetic vasococontractive pathways, 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 vasococontractive 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 of being trapped or of anger. 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 (CO2, ET) for their controls, we also note that the ET values of 38 mm Hg at 50% of maximum exercise and 37 mm Hg at maximum exercise are well below expected normal values.

The demonstration of increased minute ventilation for given minute carbon dioxide (VE/VCO2) is quite consistent with chronic hyperventilation. It is due either to reduced respiratory centre buffering or a flywheel effect which is regularly accompanied by pathways promote breathing, or both. The observation of a raised VE/VCO2 may well be the key indicator of a longer term tendency to hyperventilation, because as stated above, the FHT is a single act of hyperventilation and is dependent upon the patient's starting point in terms of other influences upon vasoconstrictive neural arousal and depletion of the body's buffering systems.

A basic question about syndrome X patients is whether their responsiveness to recognised vasococontractive influences is greater than average, just as Europeans in general are more responsive than the Japanese.

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.

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3 Lewis BI. Chronic hyperventilation syndrome. JAMA 1964;189:104-8.

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

Syndrome X—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". Our specific 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 our syndrome patients were hyperventilating inappropriately (which 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 VE/VCo2 slope but only in the presence of low arterial Pco2 (the modified alveolar gas equation states: VE = 863 Vco2/Paco2 - 1 - Vd/Vt). The normal arterial Pco2 in our patients would not lead to an increase in deadspace ventilation. We did not measure arterial Pco2 in our control patients for ethical reasons.

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

Sir,—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 other things to label this as 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,1 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,2 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.3 Several changes in morphology that were proposed, leading to rupture of the right coronary sinus into the right ventricle in young adulthood when there was a ventricular septal defect in the outlet portion of the septum.4 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.5 Indeed, during a 10 year period at the Chiang Mai hospital 32 of 36 patients undergoing surgery for a ruptured aneurysm of the sinus of Valsalva had "subarterial infundibular" ventricular septal defects.6

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 aortic valves. Prolapse of the right coronary aortic cusp results in elongation of the sinus and thinning of the sinus 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 latter complication.7

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Impending paradoxical embolism: a rare but important diagnosis

Sir,—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 al9 and 10 of 27 cases reviewed by Jungbluth et al,7 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.3

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.8 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 blood products, the morbidity, mortality, and psychological trauma associated with surgery.

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

Str,—I read Sullivan’s report with interest. The main point is that the diagnosis of a ruptured sinus of Valsalva in children by echocardiography is better. However, we classified it as a subarterial ventricular septal defect, and it was treated surgically. The etiology was thought to be congenital. It is important to understand the difference between a subarterial ventricular septal defect and a ruptured sinus of Valsalva aneurysm.

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