PostScript

Sweat testing in CF

The European Diagnostic Working Group presented comprehensive diagnostic algorithms for cystic fibrosis (CF) and confirmed the fundamental role of the sweat test for the diagnosis of CF. However, several important differences between well-accepted guidelines for sweat testing and the recommendations of the Working Group need to be discussed.

An adequate sweat sampling volume depends on the sampling area and not on the body surface area of the patient. The unit therefore has to be cited as "g/m²" sampling surface area/min rather than "g/m²" body surface area/min.

For stimulation and sampling of sweat the authors recommend only the Gibson and Cooke technique and do not even mention the widely used Macroduct collection method which is well accepted by the National Committee for Clinical Laboratory Standards (NCCLS) and UK guidelines. The authors do not give any reason for this limitation. Mastella et al. have shown an acceptable agreement between both collection systems with a mean (SD) of 2.15 (1.27) mmol/l and a mean (SD) of 2.14 (1.27) mmol/l for the Gibson and Cooke technique.

The expansion of the intermediate chloride range can more than double the number of patients who will need further diagnostic investigations. Before such a far-reaching recommendation of the expansion of the intermediate chloride range is implemented in daily routine, prospective (not only retrospective) studies are urgently needed to define the specificity and sensitivity of this modification. As we all know, even a chloride level of <30 mmol/l cannot exclude the diagnosis of CF.

Previously undiagnosed obesity hypoventilation syndrome

There are approximately 300 million obese individuals (body mass index (BMI) >30 kg/m²) or higher, worldwide, and in the UK nearly one quarter of all adults are classified as clinically obese. Obesity hypoventilation syndrome (OHS) describes a subgroup of obese individuals who develop chronic daytime hypercapnia (arterial carbon dioxide tension (Paco₂) >4 kPa) and hypoaxia (arterial oxygen tension (Pao₂) <8 kPa) in the absence of chronic obstructive pulmonary disease (COPD). Presentation is usually indolent, with symptoms arising due to hypercapnia and sustained hypoventilation (hypoxomnia, alterations in cognitive function, headache, peripheral oedema, hypertension, congestive cardiac failure).

At Southend Hospital we have noticed an increase in acute admissions in obese individuals with type II respiratory failure of initially unknown cause in whom a diagnosis of OHS was eventually made. We collected data on 11 patients (seven men) diagnosed with OHS from 1996 to 2005 from the respiratory disease register. Patients with possible overlap syndrome were excluded (smokers with forced expiratory volume in 1 s (FEV₁) <70% and mean pulmonary arterial wedge pressure >12 mmHg). The results of initial sleep studies on air were analysed. Initial management was recorded and follow-up data were reviewed regarding OHS, blood gases, long-term use of continuous positive airway pressure (CPAP) or non-invasive ventilation (NIV, using bi-level pressure support ventilation).

The mean (SD) age of the 11 patients was 59 (12) years and the mean (SD) BMI was 32.7 (16.6) kg/m² (range 20–39). Two patients were current smokers, one an ex-smoker and eight were never smokers. Seven patients were hypertensive, three were known to have hypothyroidism (two on treatment) and two had asthma. Only two cases presented before 2002 and none after 2002. Eight presented to the A&E with type II respiratory failure of unknown cause, with a mean (SD) pH of 7.25 (0.15), mean Pao₂ 11.6 (4.0) kPa and mean (SD) HCO₃⁻ 34.9 mmol/l (4.5). Three patients presented to respiratory outpatients.

All patients had increasing shortness of breath, in two the breathlessness was gradual (over 4–6 months), in six cases the breathlessness was only present in the preceding 3–4 weeks, and in three the symptoms were acute, starting only days before admission. These three patients also had evidence of a respiratory infection. All patients had a normal chest radiograph on admission. None of the patients had a diagnosis of OHS made until reviewed by a respiratory physician.

Sleep studies showed a mean (SD) apnoea/hypopnoea index score of 33 (22) (h and oxygen desaturations 39 (37)%. The range of the mean nocturnal oxygen saturation was 66–89.9%. The mean ESS was 15 on presentation (range 2–22), mean (SD) FEV₁ was 1.53 (0.52) l and mean (SD) FEV₁/FVC ratio was 77 (6%). In the eight patients presenting to A&E, six required NIV, one CPAP and one did not require intervention acutely. One patient has required treatment with NIV long-term and eight others were managed on CPAP. One patient died due to non-compliance with treatment. One has improved with weight loss alone. Only the patient with asthma has subsequently decompen-sated and developed acute type II respiratory failure.

At follow up the mean ESS was 3 (range 0–10). Blood gases on air had improved with a mean (SD) pH of 7.46 (0.10), mean (SD) Pao₂ 5.94 (1.15) kPa, mean Pao₂ 8.59 (1.38) kPa and mean HCO₃⁻ 28.6 (4.1). Decompensated OHS is often not recognised in A&E. In our study a diagnosis of OHS as the cause of respiratory failure was not appreciated until referral to a respiratory physician had been made. The presentation of OHS is very non-specific, but should be considered in obese patients who have increasing breathlessness, have never smoked, and have type II respiratory failure and a normal chest radiograph.

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References
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