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 instead of "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 Macrodust 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) difference of 0.05 (8.6) mmol/l, comparable to Denning's results which showed a mean (SD) difference between sequential Gibson-Cooke tests of -0.05 (8.6) mmol/l. Different failure rates, especially in patients under 4 months of age, should not be misused to condemn the Macrodust collection system because this problem can be overcome by experience.

The most important difference is the extension of the intermediate sweat chloride range up to 30–60 mmol/l from 40–60 mmol/l. This recommendation is based on the work of Lebecque et al. who investigated patients with sweat chloride levels of 30–60 mmol/l by extensive genetic testing and nasal potential difference measurements. Adults accounted for 30% of all patients with intermediate sweat chloride levels but were excluded from the analysis. Lebecque et al. presented 10 children with intermediate sweat chloride levels in children. Am J Respir Crit Care Med 2002; 165: 757–61.

Previously undiagnosed obesity hypventilation 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 hypventilation syndrome (OHS) describes a subgroup of obese individuals who develop chronic daytime hypopnoea (arterial oxygen tension (PaO₂) <8 kPa) and hypoxia (arterial oxygen tension (PaO₂) <8 kPa) in the absence of chronic obstructive pulmonary disease (COPD).

Presentation is usually indolent, with symptoms arising due to hypopnoea and sustained hypventilation (hypoxia, alterations in cognitive function, headache, peripheral oedema, hypertension, congestive cardiac failure). At Southend Hospital we have noticed an increase in severe obstructive sleep apnoea/hypopnoea index score of 33 (22)/h and oxygen desaturations 39 (37)/h. The mean of the range of mean nocturnal oxygen saturation was 66–89.9%. The mean ESS was 15 on presentation (range 5–22), mean (SD) FEV₁ was 1.53 (0.52) L and mean (SD) F/EV₁ 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 decompressed 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 shortness of breath, have never smoked, and have type II respiratory failure and a normal chest radiograph.