Obesity Hypoventilation Syndrome

Synonym: Pickwick syndrome

The classic features of obesity hypoventilation syndrome (OHS) are obesity and daytime hypercapnia.\(^1\) The differences between OHS and obstructive sleep apnoea (OSA) are that the former has:

- Longer and more continuous episodes of hypoventilation overnight (there may or may not be upper airway obstruction).
- Daytime hypercapnia.

Pathophysiology

Controversy surrounds the pathophysiology. Only a small proportion of morbidly obese patients have the condition, so factors other than obesity must be at work. Some authors believe that the basic problem relates to the way the ventilatory drive reacts to hypoxia and hypercapnia.\(^3\) Others consider that fat distribution, hormones and upper airway size are involved.

Upper airway obstruction is certainly known to play a prominent role. It may be associated with rapid eye movement (REM) atonia, increased fat distribution around the neck and upward displacement of the diaphragm by abdominal fat.\(^4\) Experiments with mouse models suggest that deficiency of or resistance to leptin (a substance which reduces the surface tension of lung tissue) may be involved, leading to alterations in central respiratory drive and reduced ventilatory responsiveness, permitting development of carbon dioxide retention. Changes in neuromodulators resulting from the effects of hypoxia may further exacerbate the problem by depressing arousal from sleep in the face of abnormal breathing.\(^1\)

Differentiation has been made between people who have sleep apnoea hypoventilation, and OHS in which hypoventilation is also evident whilst they are awake.\(^3\) Most patients with the syndrome also have sleep apnoea but some patients do not, suggesting that it is the obesity per se which is causing chronic hypoventilation.\(^6\)

Patients may also have concomitant features of asthma or chronic obstructive pulmonary disease (COPD).

Epidemiology

There are no figures available for incidence or prevalence, mainly because the condition has been poorly defined in the past and often confused with OSA. It has been estimated that approximately 10-20% of patients with OSA have OHS.\(^3\) Risk factors mirror that for obesity and the condition is more common in females. Tonsillar hypertrophy is an aggravating factor in children. The peak ages of presentation are 5-7 years and adolescence.\(^7\) However, increased awareness of the condition means that more and more cases are being diagnosed in adults.\(^6\)

Presentation\(^3,8\)

History

Symptoms suggestive of the diagnosis may include features of sleep apnoea (witnessed apnoea, snoring enuresis, nightmares, sleepwalking), chronic mouth breathing, daytime sleepiness, hyperactivity in children, morning headaches, decreased exercise tolerance, poor school performance, poor memory and poor concentration.

A drug history should be taken to exclude alcohol excess, sedating antihistamines and central nervous system depressants, all of which can aggravate the condition.

- The typical patient will be clinically obese, with marked fat deposition around the chin and abdomen. Thoracic kyphosis is often a feature. Leg oedema may be present.
- The head, ears, nose and throat should be examined to exclude facial dysmorphologies or ENT abnormalities that may cause upper airway obstruction, such as macroglossia, micrognathia, retrognathia or high-arched palate. The tonsils should be examined to exclude hypertrophy and the nasal passages to exclude polyps, cysts or swollen nasal turbinates.
- Examination of the chest may reveal signs of cor pulmonale (loud second heart sound, displaced cardiac impulse). Acquired pes excavatum may result from overuse of the respiratory muscles overcoming extrathoracic obstruction.

Diagnosis

OHS cannot be diagnosed on history and examination alone but requires the demonstration of daytime hypercapnia.\(^6\)

Diagnostic criteria for OHS

- Body mass index $\geq 30$ kg/m$^2$.
Differential diagnosis\(^8\)

- Sleep apnoea and other sleep-related breathing disorders (but these may co-exist).
- Prader-Willi syndrome.
- Beckwith-Wiedemann syndrome.
- Narcolepsy.
- Use/abuse of sedatives and antihistamines.
- Sleep deprivation.

Investigations\(^3,4,6\)

- Arterial blood gases - these are needed to confirm daytime hypercapnia and hypoxaemia.
- Nocturnal oximetry should be carried out to determine whether sleep apnoea is also present (about one fifth of sleep apnoeic patients will have OHS. Formal polysomnography may be required in borderline cases.
- CXR - may show chest wall deformities, or signs of cardiomegaly or congestive failure.
- Echocardiogram - may show right ventricular hypertrophy.
- ECG - arrhythmias and right bundle branch block have been recorded.
- Pulmonary function tests:
  - Flow volume loop - expiratory volume as measured by spirometry is plotted in a continuous curve against flow rate - may show a 'sawtooth' pattern associated with upper airway obstruction.\(^9\)
  - Forced vital capacity and expiratory reserve volume may be reduced and airways resistance increased.
- Overnight polysomnography may confirm hypoventilation, hypoxia and hypercapnia during sleep, especially in children and adolescents.
- FBC and TFTs should be performed to exclude anaemia and myxoedema.

Management\(^2,3,6\)

- A return to normal body weight is the mainstay of treatment. Unfortunately, although they may lose weight initially, many patients are non-compliant with dietary restriction in the long term. They are furthermore restricted from increasing their physical activity due to pulmonary symptoms. Bariatric surgery may be required in severe cases.
- Continuous positive airways pressure (CPAP) is more helpful in OSA, whereas patients with OHS usually need assisted ventilation which may need to be supplemented by oxygen.
- The inability of these patients to increase their ventilatory capacity should be borne in mind during their management (eg, when they are subjected to hospital procedures which may lead to hypercapnia).\(^10\)
- Treat any concomitant OSA, asthma or COPD as appropriate.

Complications

- Chronic hypoventilation may be associated with congestive heart failure, cor pulmonale and angina. Future research is likely to focus on the links between the syndrome and cardiovascular morbidity.\(^11\) Early epidemiological data suggest a link with coronary artery disease and stroke.\(^3\)
- Conditions associated with obesity may include arterial hypertension, diabetes mellitus, hypothyroidism, osteoarthritis, hepatic dysfunction, hyperlipidaemia, asthma and pulmonary hypertension.

Prognosis\(^3\)

Prognosis is improved by early recognition, weight loss and CPAP. Two prospective studies reported no in-hospital deaths. Retrospective studies, however, reported considerable mortality (23% in one study) in OHS patients who refused long-term CPAP.

Further reading & references


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