



# Helping the Heart: Understanding Pickwickian Syndrome

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## Adam's Case

Adam, 43, is a truck driver who had a recent motor vehicle accident and was sent to the clinic for assessment. He has:

- a ruddy complexion,
- a thick neck,
- an elevated jugular venous pressure,
- tricuspid regurgitation and
- significant peripheral edema.

He admits to falling asleep during the day and has recently noticed some leg swelling.

On exam he is morbidly obese weighing 233 kg.

His body mass index is 74 and his waist circumference is 184 cm.

His BP is 154/98 mmHg and his heart rate is 84 bpm.

**What is Adam's diagnosis?**

**For the answer, see page 21.**

Once rare, Pickwickian syndrome is increasingly common as our society grows progressively more obese. This syndrome, usually termed the obesity hypoventilation syndrome (OHS), is characterized by hypoventilation in an obese patient for which there is no other cause (Table 1).<sup>1</sup>

Pickwickian syndrome results in:

- hypoxia,
- hypercapnia,
- respiratory failure,
- pulmonary hypertension and
- right-sided congestive heart failure (*cor pulmonale*).

Left untreated, this syndrome is associated with substantial morbidity and probable early mortality.<sup>2</sup> It is important for clinicians to think of this disorder when treating obese patients.

Table 1

### Diagnostic criteria

- Body mass index > 30
- Daytime arterial carbon dioxide > 45 mmHg
- Associated sleep related breathing disorder:
  - obstructive sleep apnea-hypopnea syndrome,
  - sleep hypoventilation syndrome or
  - both
- Absence of other known causes of hypoventilation

*Once rare, Pickwickian syndrome is increasingly common as our society grows progressively more obese.*

**Q:** *What does one see clinically?*

Charles Dickens' classic novel *The Posthumous Papers of the Pickwick Club* describes the typical Pickwickian patient well; they are often morbidly obese. Patients may complain of:

- falling asleep at inappropriate times,
- morning headaches and
- mood changes.

Many patients will have a short, thick neck and a plethoric complexion (from secondary erythrocytosis). Features of *cor pulmonale* may be prominent, such as:

- elevated neck veins,
- tricuspid regurgitation,
- hepatic congestion and
- peripheral edema.

**Q:** *How does Pickwickian syndrome relate to obstructive sleep apnea?*

There is a strong association between obstructive sleep apnea (OSA) and Pickwickian syndrome. Most patients who have OHS also have OSA, but the reverse is not true. Only 10% to 15% of patients with OSA have Pickwickian syndrome.<sup>3</sup> It is not clear why some obese patients are more at risk for the development of Pickwickian syndrome.

**About the author...**

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Table 2

**Causes of hypoventilation**

Mechanical limitations:

- Underlying lung disease
- Kyphoscoliosis
- Myopathy

Neuropathic conditions:

- Diaphragmatic paralysis
- Neuropathy

Central control abnormalities:

- Severe hypothyroidism
- Cerebrovascular accident
- Central nervous system (CNS) disease
- Drugs that suppress the CNS

*More often than through medical means, bariatric surgery results in rapid, significant and sustained weight loss.*

**Q:** *How is Pickwickian syndrome diagnosed?*

Several common tests are important to order when considering Pickwickian syndrome:

- A complete blood count may reveal secondary erythrocytosis (in up to 50% of patients)
- Thyroid function, calcium, magnesium and phosphate levels may reveal factors contributing to respiratory muscle fatigue

- Arterial blood gases reveal daytime hypercapnia (arterial carbon dioxide > 45 mmHg), hypoxia and often, a compensated respiratory acidosis
- An ECG may demonstrate right atrial enlargement and right ventricular hypertrophy and strain
- An enlarged right ventricle may be seen on a chest x-ray (loss of the retrosternal airspace on lateral films)
- Pulmonary function tests are important to rule out other causes of hypoventilation, such as chronic obstructive lung disease or neuromuscular disease (Table 2)
- A formal sleep study is important to document underlying sleep disorders (apneas, hypoventilation or both)

**Q:** *How is Pickwickian syndrome treated?*

Pickwickian syndrome is treated most effectively by weight loss. Even a relatively modest weight loss of 10 kg may improve vital capacity and reduce hypercapnia. However, for most patients, this is exceedingly difficult to do.

Many experts believe that more significant weight loss is required to treat this condition and therefore recommend bariatric surgery. More often than through medical means, bariatric surgery results in rapid, significant and sustained weight loss.<sup>4</sup>

Associated sleep disturbances also need to be treated. Many patients who have both OSA and OHS can be successively treated with nocturnal continuous positive airway pressure therapy. However, some patients, especially those without OSA, will not respond to this therapy and may need noninvasive mechanical ventilation to correct the hypoventilation.<sup>5</sup> Occasionally, a tra-

**More on Adam**

Laboratory tests reveal the following:

- Blood work revealed erythrocytosis and normal electrolytes and thyroid function
- An ECG reveals right ventricular hypertrophy
- Blood gases reveal hypoxia and hypercapnia

He is sent for a formal sleep study which reveals both ventilatory obstruction and hypoventilation

He is counseled on how to lose weight and was started on:

- diuretics,
- oxygen and
- continuous positive airway pressure (CPAP).


The motor vehicle licensing board was notified of his condition.

cheostomy is required. Some patients may also benefit from supplemental oxygen therapy.

Drug therapy is important to mention. Diuretics are instrumental in treating edema. Patients should avoid respiratory depressants, such as:

- alcohol,
- sedative-hypnotics and
- narcotics.

Unfortunately, respiratory stimulants have no proven role.

Patients suffering from Pickwickian syndrome would benefit from early referral to a respirologist. 

References

1. Olson AL, Zwillich C: The obesity hypoventilation syndrome. *Am J Med* 2005; 118(9):948-56.
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3. Poulain M, Doucet M, Major GC, et al: The effect of obesity on chronic respiratory diseases: Pathophysiology and therapeutic strategies. *CMAJ* 2006; 174(9):1293-9.
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5. Berger K, Ayappa I, Chatr-Amontri B, et al: Obesity hypoventilation syndrome as a spectrum of respiratory disturbances during sleep. *Chest* 2001; 120(4):1231-8.