The Pickwickian syndrome: Special challenge for the anesthetist

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Anesthesia in markedly obese patients requires special care and is often associated with ensuing complications. Likewise, patients with pulmonary insufficiency, cor-pulmonale, and overt respiratory failure present numerous difficulties for the anesthetist.

Here, we are reporting on the case of obesity-hypoventilation or the "Pickwickian" syndrome—an uncommon but important condition in which severe obesity and respiratory failure coexist, increasing patient risk and requiring special skill in both diagnosis and management. The characteristic features of the disorder are summarized, with comments on pathophysiology, diagnostic techniques, treatment, and anesthetic considerations.

Actual case

A 29-year-old male Negro was referred to our hospital for evaluation of glycosuria. He had been obese all his adult life. His height was 5 feet 11 inches and his weight had been approximately 290 pounds for at least the last ten years. His numerous brief attempts at weight reduction with diet control had been unsuccessful.

The patient had also been suspected of having a pulmonary disorder. On one occasion four years previously, he had undergone tests which revealed mild polycythemia, a restrictive ventilatory defect as indicated on a spirogram, and arterial gases demonstrating hypoxemia and oxyhemoglobin desaturation. He was advised to lose weight, but no treatment was attempted; and he was lost to followup. A routine urinalysis of the patient made shortly before admission showed 4-plus glucose, and he was referred for further study.

The patient had no chief complaint, and in fact, denied symptoms of any kind. He stated that he never fell asleep while on duty and denied other symptoms of excessive somnolence. Past history and systems review were unremarkable; he had smoked one package of cigarettes daily for twelve years but had no respiratory complaints.

Upon physical examination, the patient (belying his stated history) was observed to fall asleep whenever the examiner ceased talking for more than a few seconds. If left undisturbed for a few minutes, he became cyanotic, his respirations became intermittent in Cheynes-Stokes pattern, and occasional twitching and jerking movements were observed in his face and extremities. His weight was 287 pounds, pulse 90, blood pressure 140/100 mmHg. The remainder of the examination disclosed no further abnormality except for marked generalized obesity. Specifically, the lungs were normal and signs of congestive heart failure were absent.
Laboratory studies included: a hematocrit of 58 vol%, a glycosuria, a diabetic-pattern glucose-tolerance test, and a normal electrocardiogram. Chest x-rays were within normal limits, except for evident obesity and symmetrical elevation of the diaphragms. Routine spirometry revealed forced vital capacity (FVC) 3.56 liters (65% of that predicted), forced expiratory volume in the first second (FEV₁) 3.02 liters (85% of the patient's FVC), and maximum mid-expiratory flow rate (MMEF) 3.68 L/sec.

To facilitate further investigation of the patient's respiratory abnormalities, a 20-gauge plastic Medi-Cut® intravenous catheter was placed in his right radial artery under local anesthesia. Arterial blood specimens could then be obtained without disturbing the patient; these were withdrawn over a three-hour period and analyzed immediately for pH and blood gases. Results are shown in Table 1.

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<th>Table 1</th>
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<td>Arterial blood gases (breathing room air)</td>
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<td>Conditions</td>
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<td>Awake, supine</td>
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<td>Asleep, supine</td>
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<td>Awake, sitting, after three minutes’ voluntary hyperventilation</td>
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The patient's glycosuria resolved and his blood glucose fell as a result of caloric restriction in the hospital, although no significant change in weight occurred. He was placed on a strict reducing diet, but failed to lose weight over several months of outpatient observation, and was thereafter lost to follow-up.*

Discussion

The name, “Pickwickian syndrome,” is taken from the Charles Dickens' character “Fat Joe” in *The Pickwick Papers.* The description of the character is an accurate account of the external manifestations of the illness. Burwell and associates, when describing the disorder clinically in 1956, first called attention to Dickens' characterization; and the combination of chronic alveolar hypoventilation and marked obesity has since been known as the Pickwickian syndrome.

Clinical features of the “classic” or original Pickwickian Syndrome are given in Table 2. All except obesity can be explained as primary or secondary effects of hypoxemia and respiratory acidosis from chronic hypoventilation. Overt right-ventricular heart failure is a latent and serious development; if it is not present, as in the case described here, the patient may volunteer no symptoms. Such individuals are often referred by concerned family, friends, or employers because of their excessive somnolence. The condition may exist undetected for years, despite ongoing chronic respiratory failure and its secondary effects.

Of the many severely obese individuals in our society, only a very few hypoventilate and develop the Pickwickian syndrome. Several attempts have been made to explain the disorder pathophysiologically. Differentiation of primary from secondary mechanisms is difficult, and research has not conclu-

Table 2
Clinical features of the Pickwickian syndrome

1. Obesity, marked
2. Somnolence
3. Twitching
4. Cyanosis
5. Periodic respiration
6. Polycythemia, secondary
7. Right ventricular hypertrophy
8. Right ventricular failure

*This particular case has been used as an example in an article on the pathophysiology and management of the Pickwickian syndrome which was submitted to *Respiratory Care.*
sively explained why the Pickwickian patient hypoventilates while an equally obese "normal" individual does not.

A fundamental factor appears to be differences in central ventilatory control. Pickwickian patients, presumably on a congenital basis, have markedly decreased ventilatory responses to hypoxia and inhaled carbon dioxide when compared to normal individuals. Their medullary respiratory centers are relatively insensitive to these stimuli, and they do not experience "air hunger" in association with increased work of breathing and other pulmonary derangements found in obesity. The result is chronic hypoventilation.

A contributing factor to hypoventilation, especially during sleep in some patients, appears to be intermittent upper airway obstruction due to infiltration of submucosal tissues with fat. Tracheostomy has abolished the defect in several reported cases.

To establish the diagnosis of the Pickwickian syndrome in a given individual, it is necessary to demonstrate: (1) chronic hypoventilation, and (2) normal underlying lung function. Hypercapnia on arterial blood gas analysis indicates alveolar under-ventilation; a concomitant normal pH is substantiation of the chronic nature of the condition. Although hypoxemia was not found in the initial arterial specimen in our patient, his elevated hematocrit (58 vol%) suggested excess red blood cell production in response to chronic hypoxemia.

Severe chronic obstructive lung disease is a common cause of chronic hypoventilation, and this must be excluded as a diagnosis if the strict criteria for the Pickwickian syndrome are to be met. Our patient's spirogram revealed a mild-to-moderate restrictive ventilatory defect, with normal unobstructed flows, consistent with the limitations of chest excursion imposed by obesity.

In a patient with the clinical features of the Pickwickian syndrome, the diagnosis may be established with more certainty if normal ventilation and oxygen transport can be shown during voluntary hyperventilation. Diagnostic blood gas analysis can be accomplished by frequent puncture of the radial artery or by insertion of a teflon catheter. After the patient has undergone three minutes of vigorous deep breathing, an arterial blood reading should demonstrate his ability to "blow off" carbon dioxide well into the hypocapnic range, with resultant increase in arterial oxygen tension. (See Table 1.)

Treatment

Weight reduction and strict bronchial toilet lead to improvement of the ventilatory status and blood gases. Treatment of the secondary medical complications, such as congestive failure, cor pulmonale, right ventricular hypertrophy, with diuretics, digitalis, and antimicrobial drugs, aids in the reversal of the cardiocirculatory abnormalities.

Many obese individuals lack the will power for weight reduction. Studies have been conducted utilizing medroxyprogesterone acetate (Provera®) or progesterone for both acute and chronic therapy in Pickwickian patients, with encouraging results. Provera® given sublingually every eight hours appears effective in improving ventilation, and reducing hypercarbia, packed cell volume, and pulmonary arterial pressure.

Anesthetic Considerations

One of the very real nightmares for the surgeon and the anesthetist is the obese patient. When presented with an extremely obese patient for an emergency procedure, the surgical team may encounter the following: (1) an extremely difficult venipuncture, (2) difficulty in airway management, and (3) the additional management of some underlying pathology.

Anesthesia for the extremely obese patient is difficult; but, for the Pickwickian patient, there are special considerations. Preoperatively, the obese patient must be carefully evaluated with regard to pulmonary function, and the underlying pathology for ascertainment
of the Pickwickian syndrome. Ideally, for elective procedures, the Pickwickian patient should be encouraged to lose weight and be treated for control of the pathophysiological conditions. Reversal of the somnolence and the polycythemia, in particular, aids to restore normal respiratory functions and to correct the hypercapnia.

Both electively and for an emergency procedure, placement of a venipuncture may be difficult; and the danger of inadvertent puncture of an artery exists. A veno-cut down should be considered to minimize risk and to ensure adequate fluid replacement during surgery.

Pharmacologically, the increase of fatty tissues greatly enhances the storage of many agents and the concomitant prolongation of their actions. A larger than normal dose of thiopental is clinically needed for induction with these patients and can result in depression of respiratory and cardiovascular systems concurrently with the strong possibility of laryngospasm. Therefore, we recommend awake oral- or nasal-intubation to ensure adequate ventilation and the prevention of depression to the cardiovascular system. Selection of an agent should be limited to one that is the least depressive to bodily functions and one that is easily reversed.

Muscle relaxation is not a problem with regard to paralysis but is a problem with regard to adequate mechanical ventilation. Because the chest is large and fat-laden, a high tidal volume (12:15 cc/kg) is required along with careful analysis of the ABG's to maintain norms. Regional anesthetic attempts would be extremely difficult due to distorted or obliterated landmarks.

Postoperatively, the somnolence and coexisting hypoventilation should be carefully considered. The Pickwickian patient does not demonstrate the "air hunger" symptom that the "normal" obese patient does because of his or her insensitivity to hypercapnia. The somnolence, coupled with the depressant effects of the anesthetic agent, can lead to an even greater alveolar hypoventilation with resultant catastrophe.

Miller and Granada reported three in-hospital mortalities related to hypoventilation in non-anesthetized patients. Incomplete reversal of muscle relaxants and recirculation of fat-stored agents are contributing factors in the immediate postoperative period; and thus, we recommend mechanical ventilation for several days postoperatively, especially in those patients undergoing upper abdominal or thoracic operations.

Conclusion

Careful anesthetic consideration for the Pickwickian patient must be accompanied by a knowledge of the amount of alveolar hypoventilation and existence of any pathophysiology. Decreased expiratory reserve volume (ERV), restrictive diaphragmatic and chest movements, hypercapnia, and the underlying pathophysiology all make the preoperative, operative, and postoperative assessment and management of the Pickwickian patient of vital concern to the anesthetist. Although rare in occurrence, the Pickwickian syndrome is also found in the obese pediatric patient.

REFERENCES


AUTHORS

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