IDIOPATHIC ALVEOLAR HYPOVENTILATION

the occurrence of complete A-V block. This patient had evidence of left axis deviation prior to the procedure. This suggested disease of the anterior superior division of the left bundle. The appearance of RBBB in this patient during right-sided catheterization left him with the left posterior inferior division of the left bundle as the only conduction pathway from the common bundle to the ventricles. Because trauma might occur to this conduction pathway when the left ventricle was entered, a standby pacing catheter was passed into the right ventricle before going on to left ventricular catheterization. Subsequently, the patient developed complete A-V block while a catheter was being passed into the aorta through the right femoral artery.

It is difficult to say why conduction failed in the posterior inferior radiations of the left bundle before the catheter entered the left ventricular cavity. Bundle branch block has been reported to occur during passage of needles or cannulae into blood vessels. This type of transient block in conduction may be due to an inappropriate vasovagal type response. Alternatively, passage of the catheter through the aorta in this patient may have been coincidental. Clinically, patients with right bundle branch block and left axis deviation frequently go on to develop transient or permanent complete heart block as they age or as the disease process effecting the RBB and the anterior radiations of the left bundle extends to jeopardize the posterior inferior radiations of the left bundle. In our patient, the posterior inferior radiations may already have been compromised, but to a lesser extent than the anterior superior radiations. Dependence of all A-V conduction on the posterior radiations, as in this case, would cause any failure of conduction through this fascicle to present as complete heart block or as the dropped beat of Mobitz type II second degree heart block. In our patient both transient complete heart block and Mobitz II occurred.

From this experience, we feel that a standby pacemaker is warranted in patients with previous left axis deviation who develop right bundle branch block during right heart catheterization. This indication for a standby pacemaker should be added to those previously accepted; i.e., permanent LBBB before right ventricular catheterization and permanent RBBB before left ventricular catheterization.

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Idiopathic Alveolar Hypoventilation

Related to Head Trauma*

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The clinical, laboratory, and postmortem findings in a patient with the idiopathic hypoventilation syndrome probably related to head trauma are described.

The following report illustrates the first case of the idiopathic hypoventilation syndrome probably related to head trauma.

CASE REPORT

A 44-year-old white man was first hospitalized at the Long Beach (California) Veterans Administration Hospital from June 1, 1965 until September 2, 1965 because of episodes of unconsciousness for 21 years. At age 23, he became unconscious following multiple blows to his head. Two weeks later, he developed an episode of unconsciousness which lasted for two minutes. These episodes gradually increased in frequency during the next 21 years, occurred from once weekly to twice daily, and lasted from 1 to 20 minutes. He developed apnea, cyanosis, loss of muscle tone, and complete loss of consciousness during these episodes; no ictal phenomena were associated. He had been hospitalized many times at different hospitals during this 21-year period with the diagnosis of post-traumatic epilepsy and had been treated unsuccessfully with diphenylhydantoin, phenobarbital, and primidone. There was no family history of any neurologic disorder.

He was 5 feet, 8 inches tall and weighed 177 pounds; his blood pressure was 140/80 mm Hg and pulse was regular at a rate of 82 beats per minute; his respiratory rate was 20 respirations per minute. He had a cyanotic appearance; physical examination was otherwise within normal limits. His hemoglobin ranged from 16.9 to 19.5 g percent;

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hematocrit ranged from 53 to 63 vol percent; white blood cell and platelet counts were normal; serum protein-bound iodine, calcium, phosphorus, sodium, potassium, and blood urea nitrogen tests were normal; spinal fluid serology was negative and protein normal; a five hour oral glucose tolerance test was normal; chest and skull roentgenograms, electrocardiograms, and electroencephalograms were normal. Pulmonary function studies on June 24, 1965, revealed no evidence of restrictive or obstructive lung disease; his tidal volume was 235 ml per breath while breathing room air, and 206 ml per breath while breathing 100 percent oxygen; minute volume was 5.57 L/min while breathing room air and 4.15 L/min while breathing 100 percent oxygen; end-expired carbon dioxide tension was 5.57 mm Hg while breathing room air, and 4.15 L/min while breathing 100 percent oxygen; minute volume was 5.57 L/min while breathing room air and 4.15 L/min while breathing 100 percent oxygen; end-expired carbon dioxide tension was 5.57 mm Hg while breathing room air, and 4.15 L/min while breathing 100 percent oxygen. While breathing room air, his arterial oxygen tension was 44 mm Hg, arterial carbon dioxide tension was 58 mm Hg, and arterial pH was 7.38. Breathing 100 percent oxygen for 12 minutes reproduced one of his apneic episodes which was treated by ventilating the patient. His arterial oxygen tension rose to 510 mm Hg, his arterial carbon dioxide tension rose to 80 mm Hg, and his arterial pH fell to 7.24 at the time of his apneic episode.

He was started on a regimen of periodic phlebotomies. He was treated for four months with ethamivan in doses from 40 to 480 mg daily, without any benefit. On March 30, 1966, his minute volume was 9.38 L/min while breathing room air, 6.10 L/min while breathing 100 percent oxygen and 9.11 L/min while breathing 2.18 percent carbon dioxide.

On May 3, 1966, he had right heart catheterization which revealed normal pressure measurements and no evidence of any shunt. On June 14, 1966, his arterial oxygen tension was 48 mm Hg, arterial carbon dioxide tension was 65 mm Hg, and arterial pH was 7.35. After voluntary hyperventilation, his arterial oxygen tension rose to 107 mm Hg, arterial carbon dioxide tension fell to 36 mm Hg, and arterial pH rose to 7.53.

On June 4, 1967, intermittent electrophrenic respiration was instituted at the Long Beach Veterans Administration Hospital in this patient by means of a catheter electrode inserted through the wall of the superior vena cava. The electrode tip was positioned so that the right phrenic nerve was stimulated through the wall of the superior vena cava. During the next 11 weeks, diaphragmatic pacing was performed twice daily for periods of 30 to 45 minutes. While this routine was continued, the patient had no further episodes of syncope. On several occasions during these 11 weeks, diaphragmatic pacing was discontinued over a weekend with recurrence of his syncopal episodes. Before electrophrenic respiration, his arterial oxygen tension was 48 mm Hg, arterial carbon dioxide tension was 65 mm Hg, and arterial pH was 7.39. After electrophrenic respiration for 30 minutes, his arterial oxygen tension rose to 86 mm Hg, arterial carbon dioxide tension fell to 38 mm Hg, and arterial pH rose to 7.49. On August 19, 1967, the catheter electrode was removed because it was infected.

On January 28, 1968, he expired at the Long Beach Veterans Administration Hospital while awaiting design of a permanent implantable diaphragmatic pacer. Microscopic sections of the brain stem, including the respiratory center in the medulla, revealed depletion and degeneration of the ganglion cells. Figure 1 illustrates degeneration and depletion of the ganglion cells in the respiratory center of the medulla.

**DISCUSSION**

To the best of our knowledge, our patient represents the first case of the idiopathic alveolar hypoventilation syndrome probably related to head trauma. Rhoads and Brody suggest that the rare syndrome of idiopathic alveolar hypoventilation with respiratory insensitivity to carbon dioxide and with voluntary hyperventilation returning arterial oxygen tension and arterial carbon dioxide tension to normal or near normal levels can be extended to include some patients with obstructive and restrictive lung disease.

Previous autopsied cases of the idiopathic alveolar hypoventilation syndrome have revealed either no histologic changes in the brain or nonspecific abnormalities.

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