Dear Editor,

Autoimmune hepatitis was first described in 1951 and in 1956 its association with antinuclear antibodies was first established. It was the first liver disease in which medical therapy actually improved survival. Autoimmune hepatitis is a relatively rare disease with a noted incidence of 160 cases per 1,000,000 in European populations. Of note, there seem to be large variations across populations with a prevalence as high as 42.9/100,000 in Alaska and 24.5/100,000 in New Zealand. There can be wide variability in the presentation of autoimmune hepatitis in regards to its histological, laboratory, and clinical features. At this point, the etiologic mechanism behind the disease remains unknown. We will present the case of a patient in the intensive care unit who required a prolonged period of mechanical ventilation secondary to autoimmune hepatitis.

A 69-year-old male presented to the intensive care unit following a mitral valve replacement and a five-vessel coronary artery bypass. He had a medical history of coronary artery disease, myocardial infarction, hypertension, diabetes mellitus type 2, cirrhosis, alcohol abuse, dyslipidemia, gastroesophageal reflux disease, glaucoma, and benign prostatic hypertrophy.

Over the ensuing 12 days, multiple attempts were made to wean the patient from mechanical ventilation. Despite the discontinuation of all sedation, the patient remained unresponsive and was unable to follow commands. As a known alcoholic with cirrhosis, ammonia levels were monitored and noted to be elevated. Ammonia levels remained elevated despite treatment with multiple therapies including lactulose, rifaximin, and neomycin. Further medical record investigation revealed the patient might have had an autoimmune disease work-up at an outside hospital. This raised concern that autoimmune hepatitis could be contributing to the elevated ammonia levels; a immunology panel was drawn, and the results showed a positive ANA and elevated anti-RNP, C-reactive protein, histone antibody and rheumatoid factor, and biopsies usually reveal small T-lymphocyte infiltration accompanied by plasma cells and occasional histiocytes [Figure 1]. The mainstay of treatment for autoimmune hepatitis is corticosteroids alone, or in conjunction with azathioprine. The patient was started on solumedrol, and within 24 h, his ammonia levels significantly decreased, and he regained consciousness and was following commands. He was successfully extubated the following morning.

Autoimmune hepatitis can be split into two types. Type 1 is characterized by the presence of anti-smooth muscle antibodies or antinuclear antibodies. It is prevalent in the adult population, while Type 2 autoimmune hepatitis is more common in children. Type 2 autoimmune hepatitis is characterized by the presence of liver–kidney microsomal antibody Type 1 or liver cytosol antibody Type 1. Most patients are female (3:1). The presence or absence of additional autoimmune markers varies from patient to patient.

Autoimmune hepatitis typically presents as acute hepatitis in approximately 25% of patients; however, it can present as fulminant hepatitis. The patient will often present with nonspecific complaints such as fatigue or arthralgia. However, patients can also present with signs of advanced liver disease such as spider angioma, GI bleeds, and jaundice. Liver enzyme elevation is often seen. Of note, elevated IgG levels are common with IgA and IgM levels remaining at normal levels.

While the cause of autoimmune hepatitis remains unknown the most prevalent hypothesis is that it develops in genetically predisposed individuals. Following exposure to triggering factors such as microbes, viruses, or other environmental factors the autoimmune attack against the liver is continued, possibly through molecular mimicry promoted by diminished control of regulatory T cells. There appears to be an association with genes in the human leukocyte antigen region of Chromosome 6 which play an essential role in the presentation of peptide antigens to CD4 T cells.

The mainstay of treatment for autoimmune hepatitis is corticosteroids, either alone, or in conjunction with azathioprine. When the response to first-line therapy has been poor, these patients have also been successfully treated with cyclosporine, tacrolimus, or mycophenolate. At this time, a targeted monoclonal therapy has not been developed for autoimmune hepatitis; only broad immune suppression has been shown to be an effect therapeutic route. These patients will typically require lifelong immune suppression.
In patients requiring mechanical ventilation approximately 26%–43% of patients fail the first weaning attempt.\textsuperscript{[5,8]} Weaning accounts for 40% of ventilator support time.\textsuperscript{[4]} Prolonged mechanical ventilation is associated with higher costs and increased complications including pneumonia, diaphragmatic atrophy, and critical illness polyneuropathy/myopathy, along with various neurologic causes, metabolic disorders, endocrine dysfunction, electrolyte disturbances, malnutrition, obesity, and anemia. Prolonged mechanical ventilation has also been associated with mortality rates as high as 25%.\textsuperscript{[8]}

Here, we report the diagnosis and treatment of a patient with autoimmune hepatitis who required prolonged postoperative ventilation.\textsuperscript{[9]} Autoimmune hepatitis is a chronic disease characterized by continuing hepatocellular inflammation and necrosis.\textsuperscript{[3]} It can present as acute or chronic hepatitis, and in rare cases, it can present as fulminant hepatic failure.\textsuperscript{[4]} This case highlights the importance of persistence in determining the cause of a patient’s prolonged ventilator dependence.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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