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CASE REPORT

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Non-Invasive Ventilatory Support in a Pediatric Patient with Guillain-Barré Syndrome in a Public Hospital in the Province of Córdoba. Case Report

Soporte ventilatorio no invasivo en un paciente pediátrico con síndrome de Guillain-Barré en un hospital público de la provincia de Córdoba. Reporte de caso

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ABSTRACT

Introduction: Patients with Guillain–Barré Syndrome (GBS) may develop acute respiratory failure as a result of muscular weakness. The optimal timing for initiating mechanical ventilation (MV) in these cases remains controversial.

Objective: To report a case of successful management using non-invasive mechanical ventilation (NIMV) in a pediatric patient with GBS treated at Hospital de Niños de la Santísima Trinidad of the Province of Córdoba.

Case description: A 6-year-old male patient presented with ocular pain, diplopia, ataxia and progressive muscle weakness, initially affecting the lower limbs and subsequently involving the upper limbs. He had a prior episode of gastroenteritis one week before symptom onset, along with specific anti-ganglioside antibodies. The medical team diagnosed an atypical presentation of GBS, characterized by rapid progression of clinical symptoms and an unusual pattern of neurological involvement. Given bibasal hypoventilation on auscultation despite preserved lung volumes on chest radiography, together with hypophonia and an ineffective cough (cough peak flow, 60 L/min), but with adequate airway protection as evidenced by effective swallowing of saliva and respiratory secretions, NIMV was initiated via an oronasal interface without supplemental oxygen. Additionally, lung volume recruitment (LVR) and mechanical insufflation-exsufflation (MIE) were performed. On day 32, the patient was discharged from the hospital, breathing spontaneously on room air.

Conclusion: This case report describes a patient with GBS who received early NIMV while maintaining adequate functional airway protection. This intervention, combined with LVR and MIE strategies, helped manage ventilatory failure and prevent the need for invasive mechanical ventilation (IMV) and contributed to a favorable clinical outcome.

Key words: Neuromuscular diseases; Guillain-Barre syndrome; noninvasive ventilation; respiratory therapy

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RESUMEN

Introducción: Los pacientes con Síndrome de Guillain-Barré (SGB) pueden desarrollar insuficiencia respiratoria aguda como consecuencia de la debilidad muscular. El momento óptimo para iniciar la ventilación mecánica (VM) en estos casos sigue siendo un tema controvertido.

Objetivo: Reportar un caso de manejo exitoso con Ventilación Mecánica No Invasiva (VMNI) en un paciente pediátrico con SGB atendido en el Hospital de Niños de la Santísima Trinidad de la Provincia de Córdoba

Presentación del caso: Paciente masculino de 6 años de edad que consultó por dolor ocular, diplopía, ataxia y debilidad muscular progresiva, que inició en los miembros inferiores y posteriormente comprometió los miembros superiores. Con un episodio de gastroenteritis una semana previa a los síntomas y resultados anticuerpos antigangliósidos específicos. El equipo médico diagnostica SGB de presentación atipica caracterizado por la rápida progresión del cuadro clínico y el patrón inusual de afectación neurológica. Ante hipoventilación bibasal auscultatoria con volumen radiográfico conservado, sumada a hipofonía y tos débil (pico de flujo tosido 60 L/min), aunque con una correcta protección de la vía aérea -evidenciada en una deglución eficaz de la saliva y las secreciones, se inició VMNI por interfase buconasal sin oxígeno suplementario. Además, se realizó reclutamiento de volumen pulmonar (RVP) y Asistencia Mecánica de la Tos (AMT). El día 32 fue dado de alta hospitalaria, respirando espontáneamente en aire ambiente.

Conclusión: Se reportó el caso de un paciente con SGB que, al conservar una adecuada protección funcional de la vía aérea, recibió de forma temprana VMNI. Esta intervención, combinada con estrategias de RVP y AMT, permitieron tratar la insuficiencia ventilatoria, evitar la VMI y facilitar una evolución clínica favorable.

Palabras clave: Enfermedades neuromusculares · síndrome de Guillain-Barré · ventilación no invasiva · terapia respiratoria

INTRODUCTION

Guillain-Barré syndrome (GBS) is an inflammatory disorder of the peripheral nervous system that, in its classic forms, is characterized by symmetric, rapidly progressive, and typically ascending muscle weakness accompanied by diminished or absent deep tendon reflexes.[1]

Several GBS subtypes have been recognized, including acute inflammatory demyelinating polyradiculoneuropathy, acute motor axonal neuropathy, and acute sensorimotor axonal neuropathy. Disease progression may be rapid, with most patients reaching peak disability within 2 weeks of symptom onset.[2,3]

Atypical GBS, in contrast, is characterized by less common manifestations, including upper extremity weakness, ptosis, neck stiffness, inability to stand because of proximal muscle weakness, headache, and dysphagia.[4]

GBS is believed to result from an abnormal immune response triggered by infection, leading to peripheral nerve injury. However, its pathogenesis is not yet fully understood. Approximately two-thirds of patients report a respiratory or gastrointestinal infection within the preceding 2 to 3 weeks. *Campylobacter jejuni* is the most frequently identified pathogen (30%).[5]

Neuromuscular disorders in children and adolescents may affect, to varying degrees, the 3 muscle groups involved in ventilation and airway protection: inspiratory muscles, expiratory muscles, and innervated bulbar muscles. Patients with GBS may develop acute respiratory failure (ARF) as a consequence of progressive weakness of the inspiratory and expiratory muscles, as well as bulbar dysfunction. The optimal timing for initiating mechanical ventilation in these cases remains controversial.[6]

To date, evidence regarding the use of NIMV for the management of respiratory failure in patients with GBS is limited, and available studies have been conducted exclusively in adult populations.[7,8] In this context, we report the successful use of NIMV in a pediatric patient with GBS treated at our institution.

This study was conducted in accordance with the Guidelines of the Declaration of Helsinki and Good Clinical Practice, with strict protection of patient confidentiality. Written informed consent for publication was obtained from the patient's parents.

CASE DESCRIPTION

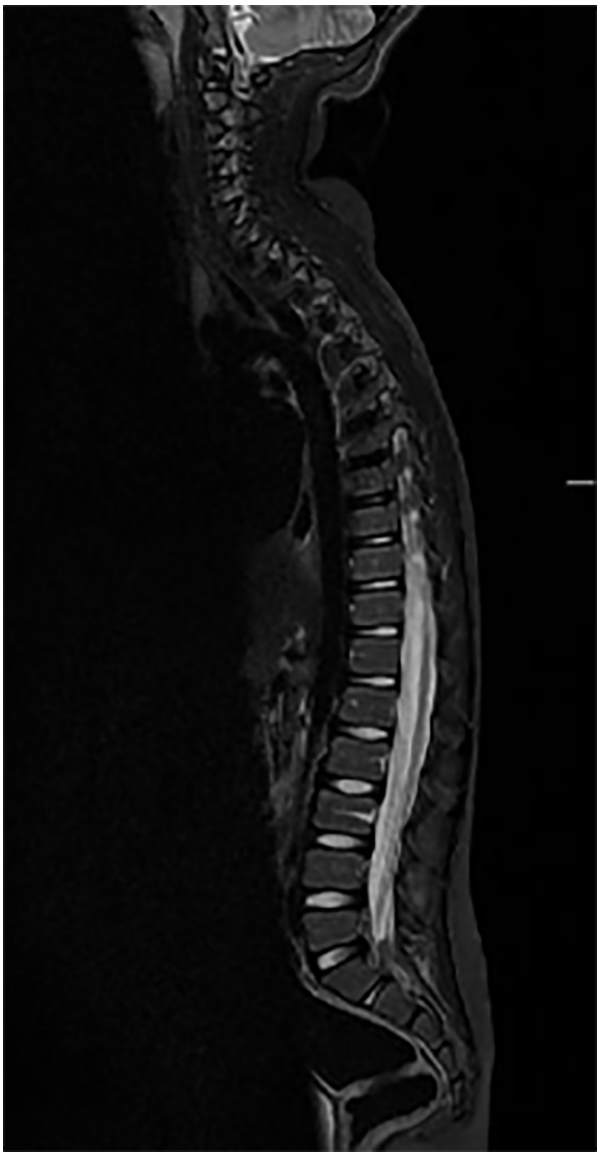
A 6-year-old boy presented to the Emergency Department of Hospital de Niños de la Santísima Trinidad, Córdoba, Argentina, on April 10, 2025. He reported ocular pain, diplopia, gait impairment, and progressive muscle weakness that initially involved the lower extremities and subsequently progressed to the upper extremities.

Upon admission to the Pediatric Intensive Care Unit (PICU), the patient was breathing spontaneously on room air and had a Glasgow Coma Scale score of 15/15. He was awake, alert, and responsive to stimuli. Deep tendon reflexes (DTR) were diminished, with apparent hyporeflexia affecting the left side of the body and flattening of the nasolabial fold. No Clonus or Babinski sign. Sensation was preserved in all four extremities. Pupils were equal, round, and reactive to light. The patient reported diplopia and exhibited marked proximal muscle weakness involving the shoulder and pelvic girdles, with inability to maintain head control. His mother reported an episode of gastroenteritis one week before symptom onset.

Lumbar puncture was performed. Serologic testing was positive for cytomegalovirus (CMV) IgM. Serum antiganglioside antibody testing yielded positive results as follows: GT1a IgG and IgM, GQ1b IgM, and in CSF: GT1a IgG and sulfatide IgG. Magnetic resonance imaging demonstrated enhancement of the cauda equina and both the anterior and posterior nerve roots (Image 1). Based on clinical findings, the medical team diagnosed an atypical presentation of GBS, characterized by rapid progression of clinical symptoms and an unusual pattern of neurological involvement.[4]

Treatment consisted of intravenous immunoglobulin (2 g/kg administered over 5 days) and analgesia. As a supportive measure, a nasogastric tube was placed, and proper positioning was ensured.

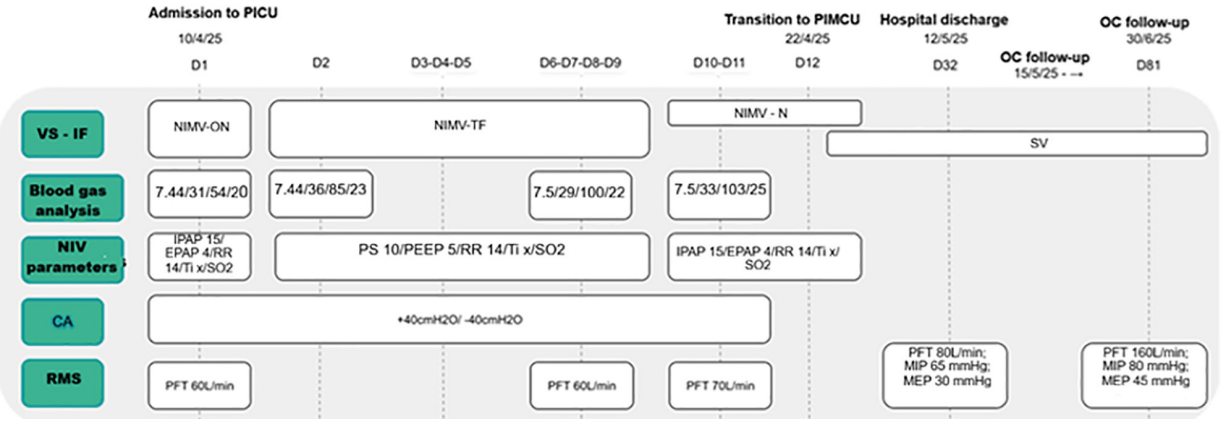
Image 1. Sagittal spine MRI (STIR sequence) showing hyperintense nerve roots



Given bibasal hypoventilation on auscultation despite preserved lung volumes on chest radiography, together with hypophonia and an ineffective cough (cough peak flow, 60 L/min), but with adequate airway protection as evidenced by effective swallowing of saliva and respiratory secretions, NIMV was initiated via an oronasal interface without supplemental oxygen. (Figure 1)

On hospital day 2, NIMV support was transitioned to a microprocessor-controlled ventilator operating in noninvasive mode with a total face

Figure 1. Timeline. Diagram illustrating the principal interventions and outcomes from hospital admission through discharge from the institution.



CA: cough assist; D: day; EPAP: expiratory positive airway pressure; IF: interface; IPAP: inspiratory positive airway pressure; N: nasal; NIMV: non-invasive mechanical ventilation; OC: outpatient clinic; ON: oronasal; PEEP: positive-end expiratory pressure; PICU: Pediatric Intensive Care Unit; PIMCU: Pediatric Intermediate Care Unit; PS: pressure support; RMS: respiratory muscle strength; *values from the latest outpatient clinic follow-up; RR: respiratory rate; SpO_2: no supplemental oxygen; SV: spontaneous ventilation; TF: total-face; Ti: inspiratory time; VS: ventilatory support

mask interface to optimize ventilatory support in response to the patient's clinical course. In addition, lung volume recruitment (LVR) was performed using a manual resuscitation bag and an inflatable mask to prevent atelectasis, facilitate secretion clearance, and improve thoracopulmonary compliance. Mechanical insufflation-exsufflation (MI-E) was also administered with pressures up to +50/−50 cm H_2O to optimize airway secretion clearance and prevent respiratory complications.[9]

On days 3, 4, and 5, functional assessments, MI-E sessions, and LVR maneuvers could not be performed because of severe radicular pain reported by the patient. Under these circumstances, any intervention that could exacerbate pain was avoided. The patient remained on NIMV with the previously established settings.

Adequate pain control was achieved on days 6 through 9. The patient continued with NIMV using a total-face interface and resumed respiratory therapy with MI-E and LVR.

On day 10, given the favorable clinical course, a well-expanded chest radiograph, and arterial blood gas values within normal limits, ventilatory support was transitioned to a continuous-flow device with a nasal interface operating in spontaneous/timed (S/T) mode.

On day 12 of PICU admission, the patient was transferred to the Pediatric Intermediate Care Unit, where progressive weaning from positive-pressure ventilatory support was continued and oral feeding was initiated. On day 32, the patient was discharged from the hospital, breathing spontaneously on room air. At the time of this report, the child remains under outpatient follow-up with neurorehabilitation, speech therapy, and respiratory therapy services.

DISCUSSION

A common clinical error is to confuse respiratory failure, typically related to abnormalities of the pulmonary parenchyma and often managed with supplemental oxygen, with ventilatory failure, which results from respiratory muscle weakness. In the latter setting, administration of supplemental oxygen without ventilatory support may mask alveolar hypoventilation and further worsen ventilatory pump failure.[9]

Respiratory impairment in NMDs is characterized by alveolar hypoventilation (resulting in hypoxemia) due to reduced ventilatory muscle pump activity despite structurally normal lungs. Supplemental oxygen does not correct alveolar hypoventilation; instead, it increases arterial oxygen tension without addressing the underlying respiratory muscle pump dysfunction. Moreover, oxygen therapy may worsen hypercapnia by blunting ventilatory drive and can mask clinical deterioration, including the presence of retained airway secretions. These patients are frequently misclassified and managed as having impaired

oxygenation due to acute respiratory failure rather than ventilatory insufficiency resulting from respiratory pump failure.[10] In our patient, early initiation of NIMV was pivotal to the favorable clinical course, providing ventilatory support that was sufficient to reduce respiratory muscle workload, optimize minute ventilation, and reverse respiratory muscle fatigue without suppressing central respiratory drive.

Acute respiratory failure develops in approximately 30% of patients with GBS, and nearly 20% require IMV.[11] In pediatric patients with GBS, IMV should be initiated at the earliest signs of respiratory muscle fatigue, before clinical decompensation occurs. Endotracheal intubation is indicated in the presence of dysphagia with salivary aspiration or ineffective airway clearance resulting from an absent or severely impaired cough.[6]

IMV is often required in GBS because recovery of respiratory muscle function is typically not expected within hours.[11] In our patient, the decision to initiate NIMV was based on the absence of impaired consciousness and bilateral facial paralysis. Although cough effectiveness was reduced, glottic closure and gag reflexes remained intact, and the patient demonstrated adequate secretion and saliva management, indicating preserved airway protection.

Consistent with our experience, a previously reported case in an adult with GBS demonstrated that NIMV could be safely used for 2 weeks in a patient who remained alert, cooperative, and capable of protecting the airway, with no evidence of dysphagia or bulbar dysfunction. This approach avoided the need for IMV and tracheostomy, and was associated with a favorable clinical outcome.7 Another study reported the use of NIMV in patients with GBS who had neither acute respiratory failure nor swallowing dysfunction and therefore maintained adequate airway protection against aspiration. In these patients, NIV avoided exposure to complications associated with endotracheal intubation, including hypoxemia, hypotension, sedation, and worsening muscle weakness.[8] In a pediatric cohort of patients with GBS, 41% required IMV, with a median duration of ventilatory support of 25 days. Among ventilated patients, 87.5% subsequently underwent tracheostomy.[12] In this context, it is noteworthy that early implementation of NIMV in our patient not only prevented progression to ventilatory failure but also avoided the need for IMV and its associated risk of tracheostomy, contributing to a favorable clinical course.

One of the key strategies in our patient was mechanical insufflation-exsufflation (MI-E). As noted by several authors, ventilatory support is often prioritized over cough-assistance techniques, despite the fact that airway clearance may be equally or even more important, particularly during the early stages of disease before ventilatory failure develops. A patient with a vital capacity of only 500 mL cannot be expected to generate an effective cough, not even with manually assisted coughing. Under normal conditions, cough produces an expiratory volume of approximately 2.3 L and peak flows ranging from 6 to 20 L/s. MI-E reproduces this physiologic mechanism by delivering a deep insufflation, typically using positive pressures of +40 to +50 cm H_2O, followed by rapid exsufflation at −40 to −50 cm H_2O.10

With respect to respiratory muscle strength assessment, a mean peak cough flow (PCF) of 179.5 ± 52.3 L/min has been reported in children younger than 10 years with neuromuscular disease (NMD).[13] These values should be interpreted cautiously, however, because normative PCF values have not yet been standardized for healthy children, adolescents, or patients with NMD in our setting.

In addition, maximal inspiratory pressure (MIP) values below 25 cm H_2O have been associated with nocturnal hypoventilation in patients with NMD. Maximal expiratory pressure (MEP) values <60 cm H_2O in individuals older than 12 years have been associated with impaired airway secretion clearance.[14]

CONCLUSIONS

This case report describes a patient with GBS who received early NIMV while maintaining adequate functional airway protection. This intervention, combined with LVR and MIE strategies, helped manage ventilatory failure, and prevent the need for IMV, and contributed to a favorable clinical outcome.

Conflict of interest

Authors have no conflicts of interest to declare.

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