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Pediatric long-term ventilation in Guillain-Barre' syndrome due to HSV1 infection: a case report

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"The baby was successfully ventilated via the tracheostomy tube with a Vivo 60 ventilator"

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Introduction

Guillain-Barre' syndrome (GBS) is the most common cause of flaccid paralysis in children and is characterized by various degrees of motor weakness, sensory abnormalities and autonomic dysfunction. In some forms, paralysis progresses very rapidly to ventilatory failure, life-threatening arrhythmias and hypertension. Infections caused by various agents have been reported to precede GBS. Associations between GBS and herpetic infections have been shown in case-control studies[1,2]. A small percentage of GBS due to herpetic infection may develop brainstem involvement with opthalmoplegia, tetraparesis and cranial nerve involvement[3,4]. Very few cases are reported in pediatric age and data about ventilatory mechanics and need of ventilation are virtually absent. Herewith, we describe a 8-months old infant who developed GBS after Herpes Simplex Virus (HSV1) infection needing mechanical ventilation both in PICU and in long term care facilities. Both parents gave the consent to publication of this report.

Case presentation

An 8-months-old boy was transferred intubated and mechanically ventilated to our tertiary teaching hospital with decreased conscious state and acute respiratory failure. His past history included varicella infection 20 days before ED admission. He had no history of prematurity, neurological illness or connatal viral infection. His recent history included coughing and fever since 24 hours with a fast decay in motility and consciousness. At admission, Glasgow Coma Scale was 3 (E1 V1 M1), in absence of deep tendon reflexes and palsy of cranial nerves. Blood gases sample revealed respiratory acidosis with pH 7.25, $paCO_2$ 54 mmHg, paO_2 55 mmHg, serum bicarbonate level 24 mEq/L, serum lactate 1 mmol/L, peripheral oxygen saturation of 85%. No hemodynamic impairment was described. Biochemistry was negative and chest x-ray showed a pulmonary infiltrate on right lung apex. The CT scan of the brain did not show acute abnormalities.

A lumbar puncture was performed which demonstrated pleyocitosis (43 cells), a protein of 40 mg/dL, glucose of 75 mg/dl, no erythrocytes and a negative Gram stain. Oligoclonal bands were present on cerebral spinal fluid and blood sample. Further serological tests, including for HIV, Polimerase Chain Reaction (PCR) for Cytomegalovirus, Varicella Zoster Virus, Herpes Simplex 2, Epstein Barr Virus, Enterovirus, Influenza Virus and Mycoplasma were negative. HSV1-PCR was found positive (40 x 103 copies). At PICU admission, he was started on intravenous Acyclovir, immunomodulant therapy (plasmapheresis and intravenous immunoglobulin).

Electromyography revealed a severe impairment in conduction both on sensitive and motor pathways bilateral both in deltoid and in bicipit femoral muscles. Electroencephalography, brain stem evoked potentials were negative. Lumbosacral MRI revealed contrast enhancement in varying degrees in the spinal nerve roots surrounding the conus medullaris and extending the length of the cauda equine, indicating radicular inflammation with anterior nerve roots enhanced more intensely according to previous pediatric report[5]. Data on respiratory mechanics, showed a low neural drive (P01=0.8), very low Maximum negative Inspiratory Pressure (MIP=-3/-10 cmH_2O), and no effective clearance of secretions due to impaired cough reflexes. The Electrical Activity of Diaphragm (Edi) was assessed with a dedicated nasogastric catheter (Maquet, Solna, Sweden). The Edi trace revealed a severe conduction impairment, that means that the child was not able to trigger any assisted breath. The child was then ventilated via a cuffed tracheostomy tube (Shiley 4) in Pressure Controlled Mode with mandatory breaths along the most critical period of the illness. During the recovery time, the Edi signal was continuously monitored and weaning from controlled ventilation was obtained by switching from Pressure Controlled Ventilation to Neurally Adjusted Ventilatory Assist (NAVA) with backup safety rate 1 month

after PICU admission (Fig. 1). Due to the persistence of low muscle strength and low endurance during illness recovery time, we choose to continue the weaning with a home care ventilator aiming to transfer the baby to the rehabilitation department.

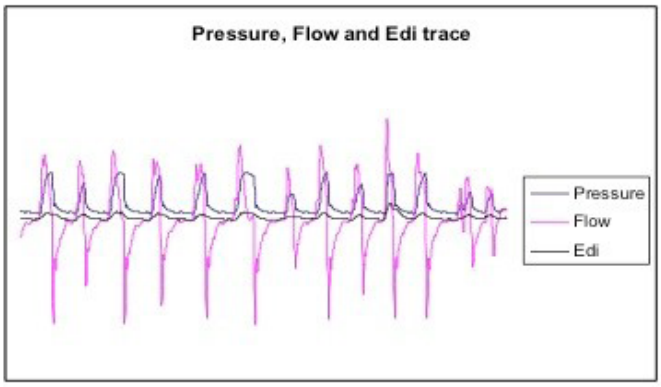

Fig 1: Pressure, Flow and Edi during weaning in NAVA

The baby was successfully ventilated via the tracheostomy tube with a Vivo 60 ventilator (Breas, Mölnlycke, Sweden), double-limb circuit in an Assisted Pressure Controlled Mode with Target Volume PCV (A+TgV) with the following settings: IPAP 15 cmH_2O, EPAP 4 cmH_2O, tidal volume expiratory 7-8 ml/kg, sensitivity trigger level 1 - most sensitive - and a backup frequency 30 breaths/min with inspiratory time of 0.6 seconds. With this setting the child was able to trigger assisted breaths without developing hypercapnia and maintaining peripheral oxygen saturation above 95% without oxygen. The weaning was then performed by reducing the backup frequency according to the improvement in Edi. He was then transferred to the rehabilitation department and fully disconnected from ventilatory support after 7 months with a complete recovery of his neurological function.

Discussion

In this paper we describe a rare presentation of HSV-1 related GBS with signs of brainstem involvement in an immunocompetent infant needing long term home mechanical ventilation. Looking at this experience, we like to address three crucial technical issues to obtain an effective ventilatory support in this particular clinical setting:

1. The trigger sensitivity
2. The measurement of the delivered tidal volume
3. The efficiency of the machine to deliver the guaranteed tidal volume (Target Volume, TgV)

Mechanical ventilation in neonates and infants with neuromuscular involvement encompasses often pressure controlled modes with backup rate and volume guaranteed function. In this setting, maintaining the spontaneous assisted breathing would be desirable, to prevent respiratory muscle atrophy due to over assistance during weaning. However, in infants it could be only a desirable end point as, in fact, even healthy infants are often unable to trigger the ventilator and to synchronize their in-expiration phases with ventilatory cycling criteria because of their characteristic breathing pattern, (i.e. fast respiratory rate, very short inspiratory time and reduced muscular strength). Consequently, wasted efforts are the most common asynchrony reported in small infants during assisted ventilation, both noninvasively and invasively delivered[6]. The low value of Maximal Inspiratory Pressure (MIP) found in this patient coupled with the severe reduction in Electrical Diaphragm Activity Signal (Edi), was consistent with a severe impairment in neuromuscular function. During the recovery period of the illness, MIP and Edi improved and the child started to trigger assisted breaths. However, in this period, many ineffective efforts were observed, due to reduction in muscle strength and inability to trigger the ventilator.

The second crucial point is the control and measurement of inspired and expired tidal volume. So, the presence of a pneumotachograph both on inspiratory and expiratory limb is crucial to control the tidal volume and to estimate the leaks.

The third crucial issue is represented by the ability of the ventilator to maintain the preset TgV without an overshooting in airway pressure to avoid barotrauma and volutrauma. We have to address, however, that this child was ventilated with a cuffed tracheostomy tube with a non-vented circuit configuration. In this configuration, it has been reported that modern ventilators are able to maintain Vt even with acute changes in respiratory impedance, both in obstructive and restrictive conditions[7]. However, the presence of an uncuffed tracheostomy tube, usually placed in smaller neonates, would have raised the point of TgV compensation[7,8]. So, we have to underline that this paper reports our experience only with double circuit non-vented configuration, and one should be careful to extend the above reported considerations to other clinical settings.

Conclusion

Looking at this clinical experience, we can conclude that:

1. small infants with GBS may completely recover from respiratory failure and be fully weaned from ventilatory support;

2. during the critical phase of the illness they often are not able to trigger assisted breaths because of reduction in muscular strength and electrical nerve transmission;

3. the recovery of neural transmission may be monitored through the Edi signal. The weaning from controlled to assisted ventilation modes could be tailored looking at improvement in electrical diaphragm conduction;

4. in this setting, the ideal home ventilator should have a very sensitive trigger, a pneumotachograph both on inspiratory and expiratory limb to measure inspired and expired tidal volume and should be able to maintain the TgV with the lowest possible overshoot in delivered pressure.

Abbreviations

GBS: Guillan Barre Syndrome, HSV1: Herpes Virus type 1, PICU: Pediatric Intensive Care Unit, ED: Emergency Department, GCS: Glasgow Coma Scale, paO_2: partial arterial oxygen tension, $paCO_2$: partial arterial carbon dioxide tension, CT: Computerized Tomography, MRI: Magnetic Resonance Imaging, MIP: Maximum Inspiratory Pressure, NAVA: Neurally Adjusted Ventilatory Assist, Edi: Electrical diaphragm Activity, TgV: Target Volume, PCV(A+TgV): Assisted Pressure Controlled Ventilation with Target Volume

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Information

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