

# A Case of Neonate with Vein of Galen Malformation Refractory to Weaning Attempts from Mechanical Ventilation Managed Successfully with Noninvasive Ventilation

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## Abstract

### Keywords

- vein of Galen malformations
- mechanical ventilation
- noninvasive ventilation

Weaning of patients with vein of Galen malformations (VOGM) from mechanical ventilation can be challenging in the postprocedure period due to underlying high-output cardiac failure and the fluid overload caused by the neurointervention procedure. We present the case of a neonate with VOGM who was refractory to multiple weaning attempts from mechanical ventilation and was successfully managed with noninvasive ventilation (NIV). NIV can be safely used as a method of weaning, post mechanical ventilation in this subset of patients. The use of NIV will facilitate successful weaning and reduce the incidence of extubation failure in neonates with VOGM in cardiac failure.

## Introduction

Patients with vein of Galen malformations (VOGMs) usually present with neurological manifestations and occasionally with severe cardiac failure, and management of these patients can be challenging, especially in the neonatal period.<sup>1</sup> We report the management of a neonate with VOGM in cardiac failure who needed emergency therapeutic intervention within the first 10 days of life after obtaining informed consent from the parents. Post intervention, the neonate was refractory to multiple attempts of conventional weaning from mechanical ventilation and was successfully weaned using a noninvasive ventilation (NIV) bridging therapy.

## Case Report

A term neonate weighing 2.6 kg delivered by cesarean section for failed induction presented with respiratory and severe cardiac failure after birth and was intubated and mechanically ventilated. Chest X-ray revealed cardiomegaly (►Fig. 1). Two-dimensional echocardiography showed severe pulmonary arterial hypertension, dilated right atrium, and right ventricle with biventricular systolic dysfunction.

A neurosonogram demonstrated features suggestive of VOGM and the neonate was referred to our institute for further management. On systemic examination, the neonate had a hyperdynamic precordium with a pansystolic murmur in the mitral area, hepatomegaly, and loud continuous bruit audible all over the cranial vault. Despite initiating the medical management, that is, injections of milrinone (0.5 µg/kg/min), isoprenaline (0.05 µg/kg/min), prostaglandin E1 (0.05 µg/kg/min), and furosemide (1 mg/kg) BD, we were unsuccessful in controlling the high-output cardiac failure.

Emergency embolization of the VOGM was undertaken to improve the cardiac function. Intraprocedural management was according to our institutional protocol. Anesthesia was maintained with end-tidal sevoflurane (1.5%), air:oxygen (50:50), and fentanyl (1 µg/kg/h) and atracurium (0.3–0.5 mg/kg/h) injections. During the procedure, the heart rate was maintained at 140 to 160 bpm, pulse oximetry was 98 to 99%, end-tidal carbon dioxide at 30 to 35 mm Hg, and the partial pressure of carbon dioxide in arterial blood [PaCO<sub>2</sub>] at 35 to 40 mm Hg. Forced-air warming system was used to maintain normothermia and arterial blood pressure was maintained in the range of 60/40 mm Hg throughout the procedure with a titrated injection of milrinone infusion (0.5 µg/kg/min)

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**Fig. 1** Chest X-ray of the neonate showing cardiomegaly.

on flow. Partial embolization of VOGM was done, wherein two out of three feeding arteries were occluded (**►Fig. 2**) and the neonate was shifted to intensive care unit for further management and kept on sedation with fentanyl (1 µg/kg/h).

Post embolization, echocardiography revealed improved biventricular function with persisting pulmonary hypertension and patient was extubated after 2 days. However, the neonate required reintubation in view of persistent hypercarbia and was taken up for follow-up angiogram and complete embolization. Postprocedure injections of noradrenaline (0.01–0.05 µg/kg/min) and milrinone (0.5 µg/kg/min) were used to maintain the blood pressure. Despite improvement in the hemodynamic profile, patient failed repeated extubation attempts and continued to require ventilator support in view of persistent hypercarbia (the partial pressure of carbon dioxide [ $\text{PCO}_2$ ] > 80). Therefore, decision was taken to give a trial extubation followed by NIV bridging support to maintain effective breathing. Nasal continuous positive airway pressure (NCPAP) was used with nasopharyngeal airway with fraction of inspired oxygen ( $\text{FiO}_2$ ) of 0.5, peak end expiratory pressure (PEEP) of 5, respiratory rate 30, and pressure control of 8 cm  $\text{H}_2\text{O}$ . The neonate was on CPAP for 2 days that was gradually weaned and discontinued and was shifted to the ward on day 4 and subsequently discharged from hospital.

## Discussion

Neonatal cardiac manifestations of VOGM can range from asymptomatic cardiomegaly to severe high-output cardiac failure and pulmonary hypertension.<sup>2,3</sup> Our patient had severe biventricular dysfunction and severe cardiac failure as 60 to 80% of the aortic blood flow was getting diverted through the low-resistance cerebral arteriovenous shunt. In our case, as the cardiac dysfunction was not responding to medical management, the patient was taken for emergency



**Fig. 2** Digital subtraction angiography of the brain shows the embolization of vein of Galen malformation (arrow).

neurointervention for embolization of the defect. Post procedure, our patient was refractory to weaning attempts mostly due to the pre-existing poor biventricular function and persistent pulmonary hypertension that led to oxygenation and ventilation defect. Our patient failed multiple weaning attempts; thus, we initiated NIV via a nasopharyngeal interface and successfully weaned the patient. In our scenario, it facilitated successful extubation and weaning by improving the cardiac function and limiting the adverse effects of hypoventilation, apnea, and postextubation atelectasis.

The initial goal of therapy in patients with VOGM presenting with failure is to arrest the congestive cardiac failure rather than to achieve complete obliteration of the arteriovenous shunt. The cardiac status transiently worsens in these patients after embolization because of sudden afterload placed on left ventricle by the closure of the low resistance, high-flow VOGM and also due to the fluid overload caused by the neurointervention procedure.<sup>3</sup> Total or near total obliteration of VOGM is avoided in single sitting because it can worsen the cardiac failure due to an acute increase in afterload caused by the removal of the low-resistance shunt. All this could attribute to difficulty in weaning and extubation of these subsets of patients in the postprocedural period.

Noninvasive ventilation in neonates has been attempted to maintain effective oxygenation and ventilation.<sup>4–6</sup> NIV helps to maintain functional residual capacity (FRC) in neonates by augmenting their spontaneous respiratory effort and minute ventilation.<sup>5,6</sup> Lung volume also influences the lung compliance in neonates, that is, smaller the lung volumes, smaller their compliance. The application of NCPAP helps in increasing the FRC, thereby improving the gas exchange resulting in better oxygenation and ventilation.<sup>6,7</sup> It is equivalent to PEEP provided through conventional ventilator without the associated adverse effects of invasive mechanical ventilation. The NCPAP has been found to be efficacious in successful weaning when used at 5 cm  $\text{H}_2\text{O}$  especially when extubation is performed within the first 14 days of life.<sup>7</sup> Nasal and nasopharyngeal interfaces have been shown to be most effective, generate least amount of airway resistance, and are minimally invasive.<sup>7</sup>

Noninvasive ventilation should be used cautiously in neonates, because if the NCPAP pressure is too high, it can result in ventilation perfusion mismatch with increasing PaCO<sub>2</sub> levels due to the hyperinflated lungs. Similarly, if low-pressure NCPAP is used, it will result in reduced lung volumes and increase in dead space ventilation thereby causing PaCO<sub>2</sub> levels to go up.<sup>7</sup> Thus, to get the optimal result with NIV in neonates and to avoid NIV-related catastrophes, a protocol-based NIV therapy should be practiced. Studies have shown that an initial CPAP of 6 cm H<sub>2</sub>O and an incremental increase by 2 cm H<sub>2</sub>O resulted in best clinical outcomes. An initial flow of 8 to 12 L/min is optimal as a high flow reduces the work of breathing (WOB), leading to a greater stability of blood pressure. Initial FiO<sub>2</sub> should be titrated to maintain PaO<sub>2</sub> values between 50 and 60 mm Hg and oxygen saturation levels between 90 and 95% to avoid retinopathy of prematurity.<sup>7</sup> Weaning failure from NIV can be avoided by reducing the CPAP by 1 cm H<sub>2</sub>O with close monitoring for any clinical deterioration. At a CPAP of 4 cm H<sub>2</sub>O with stable hemodynamics, the neonate can be safely taken off from NIV.<sup>4,7</sup> Inappropriate weaning will increase the WOB and cause rapid deterioration of respiratory function, resulting in prolonged ventilator support and convalescence.<sup>6</sup>

## Conclusion

Weaning of patients with VOGM from mechanical ventilation can be challenging in the postprocedure period due to underlying high-output cardiac failure and the fluid overload caused by the neurointervention procedure. NIV can be safely used as a method of weaning, post mechanical ventilation in this subset of patients. The use of NIV will facilitate successful weaning and reduce the incidence of extubation failure in neonates with VOGM in cardiac failure.

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## Conflict of Interest

None.

## Note

The manuscript has been read and approved by all the authors, that the requirements for authorship as stated earlier in this document have been met, and that each author believes that the manuscript represents honest work.

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