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CASE REPORT

Combined use of Neurally Adjusted Ventilatory Assist (NAVA) and Vertical Expandable Prosthetic Titanium Rib (VEPTR) in a patient with Spondylocostal dysostosis and associated bronchomalacia

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SUMMARY

Jarcho-Levin syndrome is a rare disorder characterised by defects in vertebral and costal segmentation of varying severity. Respiratory complications are the main cause of death or severe comorbidity due to a restrictive rib cage. A 3 months old infant with Spondylocostal dysostosis and associated bronchomalacia experiencing severe asynchrony during the weaning process is reported. The Neurally Adjusted Ventilatory Assist (NAVA) mode was used to improve adaptation to mechanical ventilation after Vertical Expandable Prosthetic Titanium Ribs (VEPTRs) were implanted. The synchrony achieved with the NAVA mode allowed a decrease of the sedoanalgesia he received. A follow-up CT scan showed a reduction in the volume of the posterobasal atelectasis. The evolution of this patient suggests that the combined use of VEPTR for thoracic expansion and ventilation using NAVA can favour the global improvement. This mode could be an option to consider in selected patients with difficult weaning from mechanical ventilation in paediatric intensive care units.

BACKGROUND

Spondylocostal dysostosis or Jarcho-Levin syndrome is a rare disorder characterised by defects in vertebral and costal segmentation of varying severity. It is inherited in an autosomal-recessive manner in the majority of cases, although several autosomal-dominant variants have been described.¹ Respiratory complications are the main cause of death or severe comorbidity in the first years of life. These are mainly due to a restrictive rib cage, chronic respiratory failure and frequent respiratory exacerbations.² Patients with associated tracheo-bronchomalacia have been described in the literature.³

The patients affected with malacia of the airway are especially difficult to manage as the episodes of airway collapse frequently imply the need for sedation and even muscular relaxants, consequently requiring prolonged mechanical ventilation in intensive care units (ICU).^{4 5}

The Neurally Adjusted Ventilatory Assist (NAVA) mode has been shown to offer better patient-ventilator synchrony,⁶ which was the reason for its use in this case. Ventilatory support in this mode is determined by the electrical activity of the

diaphragm (Edi) which is converted to a proportional pressure support, varying in function of the diaphragmatic electrical activity and the level of support (NAVA level) set by the clinician.

Our experience with a patient with Spondylocostal dysostosis and associated bronchomalacia is presented here, where the NAVA mode was used to improve adaptation to mechanical ventilation during the weaning process after Vertical Expandable Prosthetic Titanium Ribs (VEPTRs) were implanted.

CASE PRESENTATION

A 3-month-old infant from the UAE with a genetically-confirmed diagnosis of Jarcho-Levin syndrome and dependent on mechanical ventilation since birth was admitted to our centre for the implantation of VEPTRs. We received an intubated patient, mechanically ventilated in a volume-controlled mode, with a 0.30–0.45 fraction of inspired oxygen (FiO₂) (peak inspiratory pressures (PIP) of 23–45 cm H₂O, positive end-expiratory pressure (PEEP) of 7 cm H₂O, backup respiratory rate (RR) of 34). A chest X-ray of the thorax was performed on admission (figure 1A), and a month after the first surgery (figure 1B), showing improvement of the bilateral posterior atelectasis in the base of the lungs.

TREATMENT

Owing to the difficulty of progressing in the weaning process from mechanical ventilation after the VEPTRs were implanted to expand the thorax, a bronchoscopy was performed, showing bronchomalacia predominantly in both upper lobar bronchi. As prolonged mechanical ventilation was expected, a tracheostomy was previously performed. After an initial CT scan (figure 2A), a follow-up CT scan (figure 2B) a month after the surgery showed a resolution of the posterior-basal atelectasis, coinciding with a decrease of FiO₂ to 0.21.

Postoperatively, the patient was on the Servo-i ventilator in a volume-controlled mode and needed variable PIPs between 25 and 45 cm H₂O and PEEP values of 7–11 cm H₂O. During the attempts to wean the patient off the ventilator in the following months, he was unable to activate the



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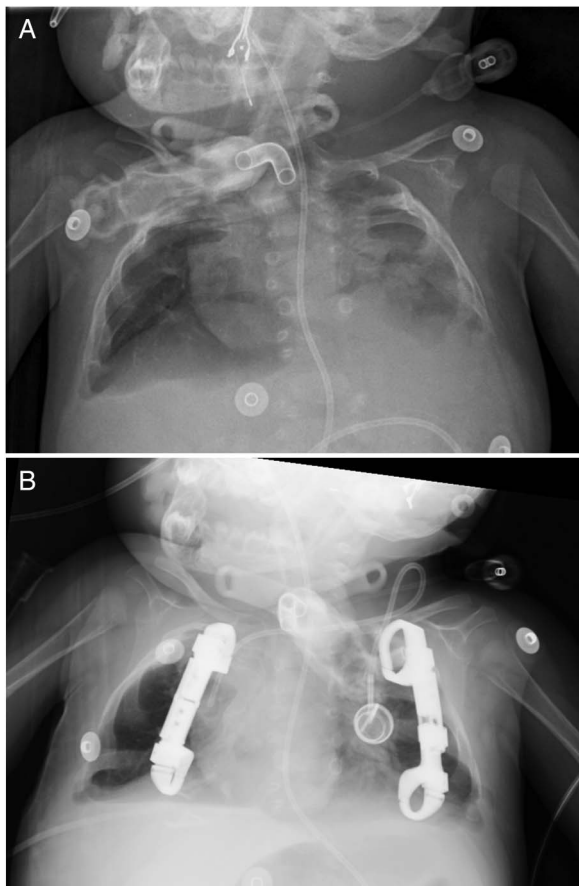


Figure 1 Chest X-ray. (A) At admission 2015, May: white left lung without mediastinal shift. Multiple vertebral anomalies. Tracheostomy. (B) 2015, June: bilateral pulmonary hyperinflation. Pleural drainage tube distal end to right paramediastinal level. Implantable venous access device (Port-a-cath), tracheostomy and right and left Vertical Expandable Prosthetic Titanium Ribs (VEPTRs).

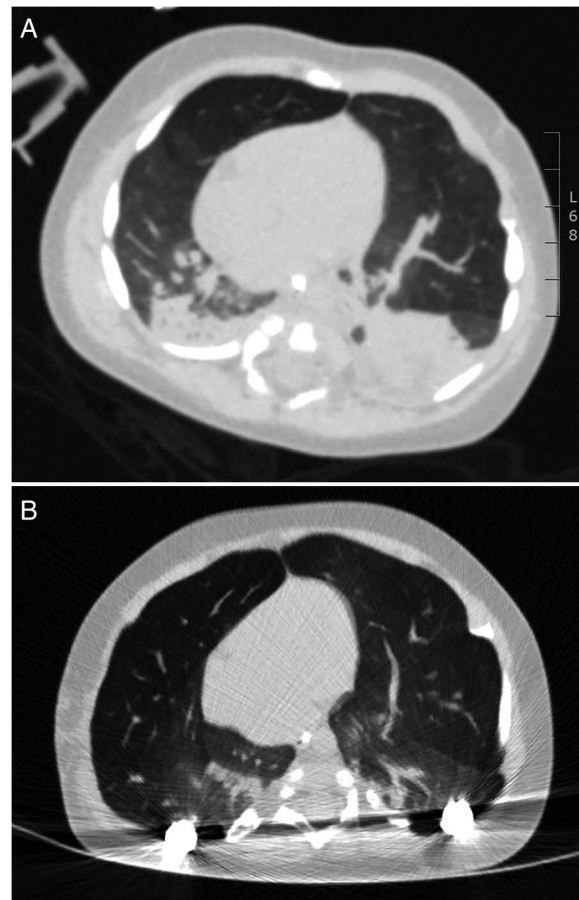


Figure 2 CT scan of the thorax. (A) At admission 2015, May: posterior-basal bilateral atelectasis. Hyperinflation with air trapping in the anterior lung parenchyma. Fusion anomalies and segmentation of all vertebral bodies. (B) After VEPTR insertion 2015, August. 1 week after starting NAVA mode: partial atelectasis in upper and lower lobes, less than prior CT. Hyperinflation with air trapping in the rest of the lung parenchyma. NAVA, Neurally Adjusted Ventilatory Assist; VEPTRs, Vertical Expandable Prosthetic Titanium Ribs.

inspiratory flow trigger and required significant sedoanalgesia to adapt to the ventilator due to repeated episodes of desaturation, on some occasions with bradycardia, associated to bronchial collapse. He continued to have asynchrony which required boluses of sedation in addition to existing medications. He initially received fentanyl and midazolam through a continuous infusion pump with progressive increase in the doses as well as continuous cisatracurium as a muscular relaxant to adapt to mechanical ventilation (figure 3). As clinical stabilisation was achieved, the muscular relaxant was withdrawn and a progressive decrease of sedation was initiated along with a change in the drug regimen. During this period, the patient developed an ocular flutter which was attributed to a pharmacological cause after neurological alterations were ruled out with normal EEG, ophthalmoscopy, metabolic workup and cranial MRI.

At 5 months of age, the NAVA mode started to improve the patient's adaptation to the ventilator through the use of a neural trigger. The NAVA catheter additionally costs around €200 per catheter; according to the manufacturer it should be changed every 5 days. It was required for 7 months; nevertheless the catheters were changed every 15 days without observing a deterioration of the Edi signal. According to the literature, Jarcho-Levin syndrome does not appear with mental retardation and life expectancy is pretty long, so ethically, we considered

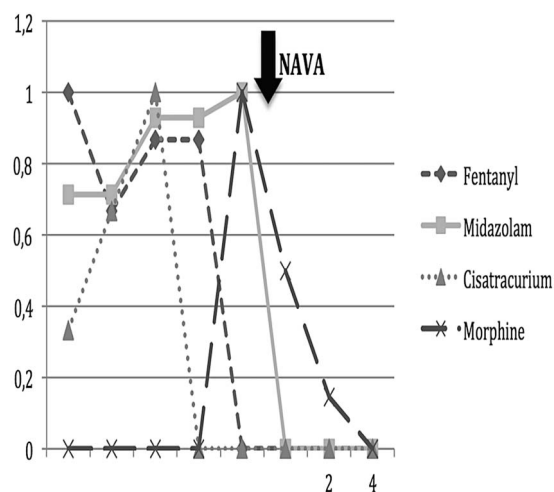


Figure 3 Sedoanalgesia administered according to time of admission. The sedoanalgesia dose administered by continuous infusion. On the vertical axis, scale of values from 0 to 1, where 1 is the maximum dose received and 0 is the suspension of the dose (equivalents: 1 Fentanyl=3 μ g/kg/hour, 1 midazolam=0.28 mg/kg/hour, 1 cisatracurium=6 μ g/kg/min, 1 morphine chloride=48 μ g/kg/hour). On the horizontal axis: weeks.

that this patient should have an opportunity to overcome his thoracic insufficiency if it were technically possible.

Tachypnoea was initially observed with a RR of 55 without other signs of increase in his work of breathing and his RR eventually returned to normal for his age (40) over the next few days. The synchrony achieved with the ventilator allowed a progressive decrease of the sedoanalgesia he received until it was completely withdrawn in 10 days; and the need for extra boluses of sedation was reduced to zero. Concurrently to the reduction of sedation, his ocular flutter disappeared and he showed significant progress in his psychomotor development.

The settings used were: NAVA level of 1 cm H₂O/μV, PEEP 11 cm H₂O, Edi trigger 0.5 μV. The patient had tidal volumes (V_T) of 6–7 mL/kg (figure 3). After 10 days on the NAVA mode, he did not require any type of sedation and the sporadic episodes of bronchial collapse were resolved with a quick increase of PEEP to 20 cm H₂O or manual ventilation with the self-inflating bag. Once he was stabilised on the NAVA mode, his PIPs oscillated between 15 and 45 cm H₂O with a NAVA level of 0.4 and Edi peaks between 15 and 100 μV.

A posterior pulmonary CT scan performed 2 months after the first surgery, 1 month after starting NAVA and prior to the first thoracic expansion, showed a significant reduction in the posterobasal atelectases previously observed (figure 2B).

OUTCOME AND FOLLOW-UP

When he turned 1 year old, after several attempts with different devices Trilogy 100 (Philips), Astral 150 (ResMed), the patient finally tolerated mechanical ventilation with a home ventilator (Monnal T-50, Air Liquide) in a volume assisted/controlled mode with the following settings: V_T 80 mL (9 mL/kg), PEEP 10 cm H₂O, flow trigger 0.5 L/min. His PIPs were around 30 cm H₂O. He did not tolerate a decrease of PEEP below 9 cm H₂O or short disconnections from the ventilator. Currently, the patient is 19 months old, is able to walk and his neurodevelopment seems to be normal.

DISCUSSION

Although the use of VEPTRs in the treatment of severe scoliosis⁷ is widely described, their use in Jarcho-Levin syndrome is more recent.^{8–9} We believe that our patient could be one of the youngest infants with VEPTRs implanted as we did not find any younger cases in the bibliography to date.

The clinical and radiological improvement of the lung parenchyma observed in this patient could be influenced by various factors in combination. On one hand, the implantation of VEPTRs and their use for thoracic expansion which result in an increase of the rib cage volume, less restriction and an improvement of the atelectasis;² on the other hand, the use of NAVA and the consequent decrease in sedation. In any case, a third CT scan prior to the first thoracic expansion continued to show resolution of the atelectasis, a phenomenon which reinforces the possible beneficial effects of combining the use of thoracic expansion with the use of NAVA. Asynchrony, a problem which is present in 25% of mechanically-ventilated patients¹⁰ is associated with increased sedation needs, worse comfort and quality of the patient's sleep, increased morbi-mortality, as well as a longer time on mechanical ventilation and length of stay in the ICU.

Our patient presented with severe inspiratory asynchrony with episodes of airway collapse which finally led to a need for increased sedation various times a day. The option used to improve this type of asynchrony was ventilation with the NAVA mode together with an elevated PEEP level. The decrease of

sedation until it was withdrawn was, we believe, related to the change in ventilation mode.

It has been shown that sedation results in atelectasis of the posterior lung. The NAVA accompanied by PEEP in our case allowed us to remove sedation with consequent resolution of the atelectasis.¹¹

Learning points

- ▶ The better patient–ventilator synchrony which allowed weaning of sedation in our case suggests that this mode could be an option to consider in selected patients with difficult weaning from mechanical ventilation due to tracheobronchomalacia in paediatric intensive care units.
- ▶ The evolution of this patient suggests that the combined use of Vertical Expandable Prosthetic Titanium Ribs technology for thoracic expansion and ventilation using Neurally Adjusted Ventilatory Assist can favour the global improvement of a highly-complex patient.

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Contributors MP-O conceived the case report and contributed to writing the manuscript. AV and NA were involved in acquisition of data and equally contributed to writing the manuscript. FJC contributed to writing the manuscript. All the authors read, reviewed and approved the final version of this case report.

Competing interests Although MP-O has been speaker for MAQUET and our institution received disposable material 6 years ago to start using this technique, we honestly consider that we are not promoting a specific product in order to favour commercial interests of the company. So, we declare this potential conflict of interest, nothing to hide, but we believe there is no a real conflict of the interest of the authors.

Patient consent Obtained.

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REFERENCES

- 1 Turnpenny PD, Young E. ICVS (International Consortium for Vertebral Anomalies and Scoliosis). Spondylocostal dysostosis, autosomal recessive. In: Pagon RA, Adam MP, Ardinger HH, et al, eds. *GeneReviews*. Seattle, WA: University of Washington, 2009:1993–2017. (Updated 17 Jan 2013).
- 2 Karlin JG, Roth MK, Patil V, et al. Management of thoracic insufficiency syndrome in patients with Jarcho-Levin syndrome using VEPTRs (vertical expandable prosthetic titanium ribs). *J Bone Joint Surg Am* 2014;96:e181.
- 3 Schulman M, Gonzalez MT, Bye MR. Airway abnormalities in Jarcho-Levin syndrome: a report of two cases. *J Med Genet* 1993;30:875–6.
- 4 Murgu S, Colt H. Tracheobronchomalacia and excessive dynamic airway collapse. *Clin Chest Med* 2013;34:527–55.
- 5 Leung L, Chung PH, Wong KK, et al. Management of tracheobronchial obstruction in infants using metallic stents: long-term outcome. *Pediatr Surg Int* 2015;31:249–54.
- 6 Ducharme-Crevier L, Beck J, Essouri S, et al. Neurally adjusted ventilatory assist (NAVA) allows patient-ventilator synchrony during pediatric noninvasive ventilation: a crossover physiological study. *Crit Care* 2015;19:44.
- 7 Balioglu MB, Albayrak A, Akman YE, et al. The effect of vertical expandable prosthetic titanium rib on growth in congenital scoliosis. *J Craniovertebr Junction Spine* 2015;6:200–5.

- 8 Mayer O, Campbell R, Cahill P, *et al.* Thoracic insufficiency syndrome. *Curr Probl Pediatr Adolesc Healthcare* 2016;46:72–97.
- 9 Ramirez N, Flynn JM, Emans JB, *et al.* Vertical expandable prosthetic titanium rib as treatment of thoracic insufficiency syndrome in spondylocostal dysplasia. *J Pediatr Orthop* 2010;30:521–6.
- 10 Verbrugge W, Jorens PG. Neurally adjusted ventilatory assist: a ventilation tool or a ventilation toy? *Respir Care* 2011;56:327–35.
- 11 Putensen C, Muders T, Varelmann D, *et al.* The impact of spontaneous breathing during mechanical ventilation. *Curr Opin Crit Care* 2006;12:13–18.

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