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Pulmonary hypoplasia and congenital dextrocardia presenting with hypercapnia in advanced age

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This is the case of a 80-year-old woman with a past medical history of congenital pulmonary hypoplasia, dextrocardia and bronchiectasis (Fig. 1 A-B), severe pulmonary hypertension, and severe mixed ventilatory failure. Six years ago, she initiated home noninvasive ventilation (NIV) for chronic hypercapnia and nocturnal hypoventilation. Optimal compliance and adherence were achieved through telemedicine. Over the past year, the patient had multiple hospital admissions for non-infectious respiratory and cardiac decompensation, with an increase in his baseline hypercapnia (67 mmHg) and hypoxemia. Despite various adjustments to her ventilatory parameters during several visits, including an increase in nocturnal O_2 to 2 litres, there was no improvement in hypercapnia and respiratory failure. We decided to perform a nocturnal polysomnography with manual titration of NIV and transcutaneous capnography, which showed a baseline O_2 saturation of 86-88%, asynchronies, flow limitation and positional snoring in the left lateral decubitus position. When the patient was changed to the right lateral decubitus position, the obstructive events and hypoventilation improved significantly with an increase in oxygen to 3 L/min (Fig.1 C). These findings demonstrated a positional component associated with the patient’s underlying congenital pathology, leading to sleep-related hypoventilation. Daytime blood gas analysis revealed a marked reduction in hypercapnia after hypoxemia was corrected. Postural interventions and continued use of her NIV were recommended.

Patient consent statement

Informed consent was requested prior to publication, which was given verbally and recorded in the patient's past medical history.

Artificial Intelligence Involvement

The author declares that he has not used any type of generative artificial intelligence for the drafting of this manuscript, nor for the creation of images, graphics, tables, or their corresponding captions.

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Authors' contribution

MMV conceptualized the case, collected clinical data, adjusted ventilatory parameters, and drafted and reviewed the manuscript.
ML contributed to the literature review, diagnostic reasoning, and manuscript drafting.
AMP conducted follow-up visits, reviewed complementary investigations, and adjusted ventilatory parameters.
All authors reviewed and approved the final version of the manuscript.

Conflicts of Interest

None declared.

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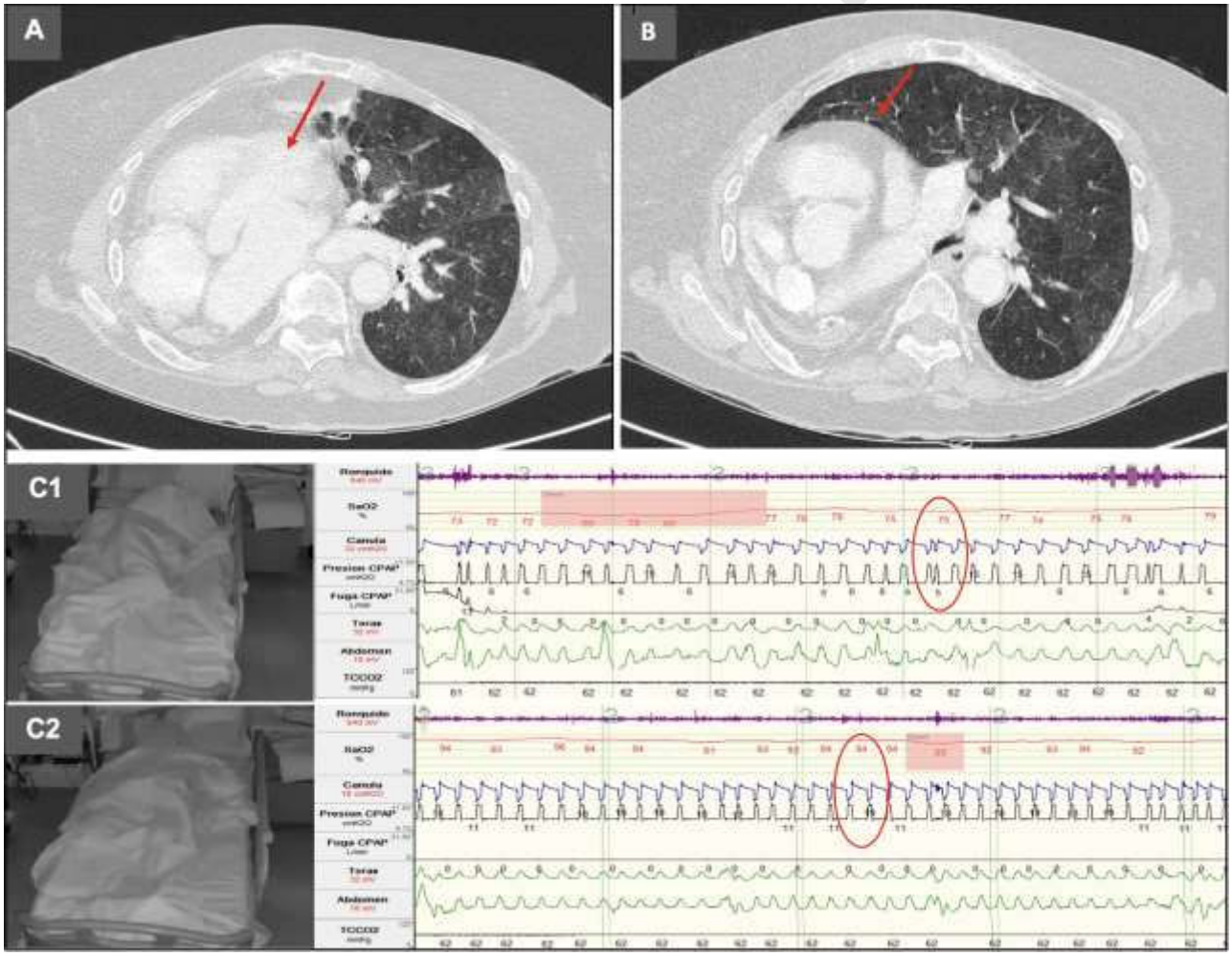

Figure 1. (A) Chest computed tomography demonstrating congenital dextrocardia (arrow). (B) Congenital pulmonary hypoplasia (arrow). (C1) NIV titration in the left lateral decubitus position showing ventilatory asynchronies and hypocapnia (oval). (C2) NIV titration in the right lateral decubitus position with resolution of asynchronies and improved oxygen saturation (oval).