

Helmet-delivered heliox-CPAP in severe upper airway obstruction caused by PHACES syndrome

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Keywords:	heliox, helmet, non-invasive ventilation, PHACES syndrome, airway obstruction

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**Helmet-delivered heliox-CPAP in severe upper airway obstruction
caused by PHACES syndrome**

Vivanco-Allende A. MD, Mayordomo-Colunga J. MD, Coca-Pelaz A. MD, Rey C.
MD, Medina A. MD

Correspondence author:

Ana Vivanco Allende

c/ Valdés nº10 2ºE, 33012, Oviedo (Asturias)

Spain

Telephone number: +34-670716926

Email address: anaviall@hotmail.com

ABBREVIATED TITLE:

Helmet-delivered heliox-CPAP in airway obstruction

SUMMARY:

We present the case of a 4-month-old girl with PHACES syndrome and severe upper respiratory airway obstruction secondary to multiple subglottic and tracheal haemangiomas effectively treated with heliox-CPAP delivered by helmet (HH-CPAP).

KEY WORDS:

Heliox; helmet; non-invasive ventilation; PHACES syndrome; airway obstruction.

For Peer Review

INTRODUCTION

Heliox, noninvasive ventilation (NIV)¹ and combined heliox and NIV therapy^{2,3} have shown effectiveness in the treatment of paediatric respiratory disease.

A relatively new interface (helmet) has been used in ARF adult patients with promising results^{4,5,6}. In paediatric patients, helmet-delivered CPAP has been used in preterm neonates⁷, children with hypoxaemic ARF⁸, acute bronchiolitis⁴ and leukaemia⁹.

We report the case of an infant with acute upper respiratory airway obstruction caused by multiple haemangiomas effectively treated with heliox-CPAP delivered by helmet (HH-CPAP).

CASE REPORT

A 4-month-old girl was brought to the paediatric emergency department because of increased work of breathing and severe stridor in the last hour.

She had been diagnosed with PHACES syndrome in neonatal period due to coexistence of segmental facial haemangioma, persistence of trigeminal artery, vertebral artery hypoplasia, microphthalmos and morning glory optic disk anomaly. She was being treated with propranolol (2 mg/kg/day). She had had two episodes of upper airway obstruction at 1 and 3 months of age, treated with heliox-CPAP administered with nasal cannulae and intravenous steroids.

Initial treatment at the paediatric emergency room was nebulized epinephrine, intravenous methylprednisolone and inhaled heliox, but patient's clinical condition worsened. The infant showed toxic appearance, severely increased work of breathing, slight cyanosis and stridor (Taussig score: 9 points). Her heart rate was 185 beats/minute and respiratory rate 28 breaths/minute. Transcutaneous oxygen saturation could not be determined. Transferred to our paediatric intensive care unit, HH-CPAP therapy was assayed (Figure 1A), with the medical team ready for an immediate intubation. A dramatic improvement in respiratory distress was noted (Taussig score: 3 points). HH-CPAP therapy could be stopped 80 hours after admission, but heliox therapy at 6 lpm through nasal cannulae was needed for 9 days.

A fibrobronchoscopy revealed multiple subglottic and tracheal haemangiomas (Figure 1B). She continued treatment with propranolol at higher doses (3 mg/kg/d) and methylprednisolone (1 mg/kg/d). Twenty days after admission she was discharged with no stridor, nor increased work of breathing.

DISCUSSION

PHACES syndrome involves a segmental facial haemangioma of infancy (H) accompanied by at least one of the following: posterior fossa malformation (P), arteriovenous malformation (A), cardiovascular anomaly (C), eye abnormality (E) or ventral defect including sternal clefting or supraumbilical raphe (S)¹⁰. Airway haemangiomas must be suspected in infants with facial haemangioma and upper airway obstruction episodes.

To our knowledge, there are no other reported cases about severe upper airway obstruction treated successfully with HH-CPAP avoiding tracheal intubation.

Heliox has been promoted as adjunctive therapy in paediatric severe upper airway obstruction^{1,11,12}. Due to its less density comparing to air or oxygen, it provides more laminar flow in obstructed airways. Furthermore, heliox improves gas exchange and aerosol deposition and decreases work of breathing¹³. Combined therapy with CPAP has also been proven effective³; while heliox contributes with the benefits listed before, NIV adds a decrease in the workload on the inspiratory muscles, avoids airway collapse and promotes heliox distribution^{3,4,14}.

Because infants are commonly nose breathers, nasal mask and nasal prongs are the most commonly used interfaces^{10,15,16}. Helmet is postulated to be a relatively new interface that has several clinical advantages in this age group as ease of use, absence of air leakage resulting from mouth opening and a more stable fixation system that could reduce cutaneous and skull bone lesions. CPAP delivered by helmet appears to be a viable method of supporting the breathing of hypoxemic infants⁴.

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37 Ana Vivanco Allende

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40 c/ Valdés nº10 2ºE, 33012, Oviedo (Asturias)

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47 Telephone number: +34-670716926

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37 Ana Vivanco Allende
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47 Telephone number: +34-670716926
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INTRODUCTION

Heliox, noninvasive ventilation (NIV)¹ and combined heliox and NIV therapy^{2,3} have shown effectiveness in the treatment of paediatric respiratory disease.

A relatively new interface (helmet) has been used in ARF adult patients with promising results^{4,5,6}. In paediatric patients, helmet-delivered CPAP has been used in preterm neonates⁷, children with hypoxaemic ARF⁸, acute bronchiolitis⁴ and leukaemia⁹.

We report the case of an infant with acute upper respiratory airway obstruction caused by multiple haemangiomas effectively treated with heliox-CPAP delivered by helmet (HH-CPAP).

CASE REPORT

A 4-month-old girl was brought to the paediatric emergency department because of increased work of breathing and severe stridor in the last hour.

She had been diagnosed with PHACES syndrome in neonatal period due to coexistence of segmental facial haemangioma, persistence of trigeminal artery, vertebral artery hypoplasia, microphthalmos and morning glory optic disk anomaly. She was being treated with propranolol (2 mg/kg/day). She had had two episodes of upper airway obstruction at 1 and 3 months of age, treated with heliox-CPAP administered with nasal cannulae and intravenous steroids.

Initial treatment at the paediatric emergency room was nebulized epinephrine, intravenous methylprednisolone and inhaled heliox **without CPAP**, but patient's clinical condition worsened. The infant showed toxic appearance, severely increased work of breathing, slight cyanosis and stridor (Taussig score: 9 points). Her heart rate was 185 beats/minute and respiratory rate 28 breaths/minute. Transcutaneous oxygen saturation could not be determined. Transferred to our paediatric intensive care unit, HH-CPAP therapy (**helmet served by CaStar; Starmed, Mirandola, Italy**) was assayed (Figure 1A), with the medical team ready for an immediate intubation. A dramatic improvement in respiratory distress was noted (Taussig score: 3 points). HH-CPAP therapy could be stopped 80 hours after admission, but heliox therapy at 6 lpm through nasal cannulae was needed for 9 days.

A fibrobronchoscopy revealed multiple subglottic and tracheal haemangiomas (Figure 1B). She continued treatment with propranolol at higher doses (3 mg/kg/d) and methylprednisolone (1 mg/kg/d). Twenty days after admission she was discharged with no stridor, nor increased work of breathing.

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DISCUSSION

PHACES syndrome involves a segmental facial haemangioma of infancy (H) accompanied by at least one of the following: posterior fossa malformation (P), arteriovenous malformation (A), cardiovascular anomaly (C), eye abnormality (E) or ventral defect including sternal clefting or supraumbilical raphe (S)¹⁰. Airway haemangiomas must be suspected in infants with facial haemangioma and upper airway obstruction episodes.

To our knowledge, there are no other reported cases about severe upper airway obstruction treated successfully with HH-CPAP avoiding tracheal intubation.

Heliox has been promoted as adjunctive therapy in paediatric severe upper airway obstruction^{1,11,12}. Due to its less density comparing to air or oxygen, it provides more laminar flow in obstructed airways. Furthermore, heliox improves gas exchange and aerosol deposition and decreases work of breathing¹³. Combined therapy with CPAP has also been proven effective³; while heliox contributes with the benefits listed before, NIV adds a decrease in the workload on the inspiratory muscles, avoids airway collapse and promotes heliox distribution^{3,4,14}.

Because infants are commonly nose breathers, nasal mask and nasal prongs are the most commonly used interfaces^{10,15,16}. Helmet is postulated to be a relatively new interface that has several clinical advantages in this age group as ease of use, absence of air leakage resulting from mouth opening and a more stable fixation system that could reduce cutaneous and skull bone lesions. CPAP delivered by helmet appears to be a viable method of supporting the breathing of hypoxemic infants⁴.

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3 **CONCLUSION**
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6 In order to avoid endotracheal intubation we propose HH-CPAP for the
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8 management of severe upper airway obstruction in unresponsive to standard therapy
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7 **Figure 1:** Composition of images showing: A) HH-CPAP therapy, after initial
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9 true vocal cords reducing airway diameter.
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3 Comment 1:
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5 It is mentioned that the patient received nasal CPAP with heliox successfully twice
6 before the current episode. It is mentioned that the child was treated unsuccessfully
7 with inhaled heliox prior to the helmet CPAP therapy: was he on nasal CPAP with
8 heliox again, or only inhaled heliox without CPAP?
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13 Response 1:

14 Thank you so much for your comment.

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17 Initially, the infant was unsuccessfully treated with inhaled heliox, without CPAP (added
18 to the manuscript).
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22 Comment 2:
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24 Please give an address where (or if) this device can be obtained
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28 Response 2:

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30 CaStar; Starmed, Mirandola, Italy (added to the manuscript).
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34 Comment 3:
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36 Was the consent of the parents obtained and the experimental nature of this device
37 explained? This should be declared in the paper, if it was done.
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41 Response 3:
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44 The use of this device is not experimental in our unit as we are employing it in infants
45 with bronchiolitis requiring CPAP support (Mayordomo-Colunga J, Medina A, Rey C,
46 Concha A, Los Arcos M, Menéndez S. Helmet-delivered continuous positive airway
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48 308-11). As a result, no special consent was obtained.
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