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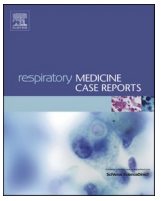

Case Report

Heliox in the management of respiratory failure in a Morquio A syndrome patient with trachea narrowing

Minghan Shi [a,b,*], Birubi Biman [a,b]

[a] Thunder Bay Regional Health Sciences Centre, 980 Oliver Rd, Thunder Bay, Ontario, P7B 6V4, Canada
[b] NOSM University, 955 Oliver Rd, Thunder Bay, Ontario, P7B 5E1, Canada

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ABSTRACT

Morquio A Syndrome is a mucopolysaccharide deposition disease where patients can develop respiratory failure due to airway narrowing by polysaccharide deposition, anatomical distortion and compression of the airway. Definitive management with surgery is high risk, only performed in the highly specialized centers, and by the time they develop respiratory failure they may not be a candidate for surgery due to difficulty intubation. Consequently, they often end up with long term BiPAP dependent and suffer from side effects of abdominal, facial pain and depression. Heliox is low density gas mixture which can facilitate oxygen delivery in narrowed airway. Here we report a case of Morquio A syndrome patient with 24-h BiPAP dependence and successfully treated with Heliox and weaned off BiPAP for 4 hours a day with improved quality of life.

1. Background

Morquio Syndrome A or Mucopolysaccharidosis type IV A is a rare autosomal recessive mucopolysaccharide deposition disorder with a birth prevalence of between 1 per 71,000 to 500,000 live births [1,2], where the body builds up glycosaminoglycans (GAGs) leading to multiple organs dysfunction [3]. It is caused by a deficiency of galactosamine-6-sulfatase (GALNS) which leads to catabolism, accumulation, and deposition of GAGs in multiple systems, including respiratory, cardiovascular systems, etc. [4,5] Respiratory failure is the main cause of death in this population [6,7].

Airway narrowing, caused by mucopolysaccharide deposition and morphological changes of the thoracic outlet, is one of the main causes of respiration failure in this population. In severe cases, patients are often dependent on ventilators which significantly reduces their quality of life. Managements of airway narrowing with stents, tracheostomy [8,9], thermoplasty has been attempted but only in experienced centre with high risk of failure due to increased perioperative risks and difficult intubation [10]. Heliox is a gas mixture of helium and oxygen. Its low density and propensity towards laminar flow reduce the airway resistance. We here report the application of Heliox in the management of this population as an alternative medical management of airway narrowing in this rare genetic disease.

2. Case presentation

A short statue female in her 40s admitted for acute respiratory distress and subsequently treated with 24-h bilevel positive airway pressure (BiPAP) having difficulty weaning it off.

* Corresponding author. Thunder Bay Regional Health Sciences Centre, 980 Oliver Rd, Thunder Bay, Ontario, P7B 6V4, Canada.
E-mail address: mshi@nosm.ca (M. Shi).

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Her past medical history includes Morquio A syndrome with several surgeries and complications including spinal dysplasia and lumbar decompression, cervical spinal fusion and decompression, scoliosis, obstructive sleep apnea (OSA) on night BiPAP and previous history of difficulty intubation and deferred tracheostomy in her 20's. She does not smoke cigarette nor use recreational drugs.

She initially developed sudden onset dyspnea on arrival of a flight. Her family denied fever, running nose, cough, sputum, or chest pain. She saturated at 92 % on 15 L/min of O_2 while getting manually ventilated, with a respiratory rate of 30 per minute, blood pressure of 139/116 mmHg, temperature of 36.1 °C. She had altered mental status, central cyanosis with decreased chest wall mobility. Lung auscultation showed decreased breathing sound bilaterally, no wheezes, rhonchi, or crackles. Normal heart sounds without murmurs. JVP normal, no lower extremities edema. Initial investigations showed respiratory acidosis with normal naso-pharyngeal swab for viral PCR, normal leukocyte count, unremarkable chest x ray and CT pulmonary angiogram but significant trachea narrowing. She was diagnosed with hypercapneic hypoxemic respiratory failure with unknown triggers and subsequently treated with antibiotics and respiratory support with BiPAP and stablized on 24-h BiPAP with the following parameters: IPAP: 20 mmH2O, EPAP: 15 mmH2O and O2 flow rate of 5L/min. However, they were not able to wean her off the BiPAP and experienced abdominal distention and facial pain, for which she was receiving hydromorphone. We were consulted at that time because of unable to wean off BiPAP.

3. Vitals and physical examination

Temperature 36.2 °C, saturation at 96 % with BiPAP and 5L/min O2, respiratory rate 16 per minute, blood pressure 108/76 mmHg, pulse 60 per minute. Short statue, short neck, decreased neck mobility, scoliosis, prominent chest wall. Alert and oriented. Lung auscultation showed good breathing sound bilaterally, no wheezes, rales, or crackles. Heart sounds were normal, no murmurs. Abdomen was distended and generalized tenderness on palpation.

3.1. Investigations

Venous blood gas: pH 7.34, pCO2: 44 mmHg, HCO3: 24 mmol/L. Normal leukocyte count. Repeat CT chest redemonstration of significant trachea structure changes with narrowing and possible of thyroid gland compression at the thoracic outlet segment.

3.2. Diagnosis

Hypercapneic hypoxemic respiratory failure in the setting of Morquio A syndrome with significant trachea narrowing caused by query trachobranchiomalacia and/or thoracic outlet organs compression, OSA and restrictive thoracic wall.

3.3. Treatment

We reviewed the case with ENT regarding possibility airway management including stenting and tracheostomy. Due to anatomical difficulties for intubation and poor respiratory reserve, the patient was determined not a candidate for these procedures. We decided to have a trial of three days of Heliox 80/20, 2–4 hours per day due to significant trachea narrowing.

Table 1
Respiratory System Involvement in Morquio syndrome [13].

Upper Airway
Upper airway obstruction, not specified [14–17]
Macroglossia, adeno-tonsillar hypertrophy, pharyngeal narrowing [17,18]
Limited mouth opening [19]
Laryngeal airway narrowing and laryngomalacia [17,20,21]
Obstructive sleep apnea [16,22–24]
Decreased mobility of the neck [17]
Lower Airway obstruction
Subglottic, trachea stenosis (tracheomalacia, deposition, short neck leads to tortuous airway, Tracheal distortion) [8,9,15–17,20,25]
Bronchus stenosis (bronchomalacia, deposition) [11,17,20,21,26]
Bronchitis, pneumonia (Thickened secretions) [15,16]
Parenchymal
Interstitial markings, unknown significance [16]
Thoracic cage
Restrictive thoracic cage (chests wall changes due to vertebral-column abnormalities, kyphoscoliosis) [14–16,23,27,28]
Obesity [23]
Neuromuscular
Muscles weakness or paralysis caused by spinal cord compression [16,23,29]

3.4. Outcome and follow-up

Immediately after the three days of Heliox trial, she was able to wean off Bipap for 4 hours per day with 30 % FiO2 high flow nasal canal support. Her abdomen was less distended which helped for her abdominal pain and reduced pain medications. However, due to limited storage and access to Heliox in our centre, we had to discontinue Heliox, but she remained off BiPAP for couple of hours per day.

4. Discussion

In Morquio A syndrome, the causes of airway obstruction are multifactorial. The mucopolysaccharide deposition in trachea wall can lead to trachea narrowing. The disproportional development of the organs and chest wall leads to internal organs distortion and compromises the airway in the space limited thoracic outlet causing obstruction. At the same time, chest wall deformity including scoliosis restricted the movement of chest wall causing restrictive disease. The causes of respiratory failure are summarized in Table 1 based on previous case reports and review articles. I suspect the patient developed Saber-sheath type trachobranchiomalacia (TBM) due to bilateral softening of the cartilaginous wall and narrowing the transverse diameter [11]. Although the pathological progression of TBM is gradual, once the critical level of compromise is reached or precipitated by certain risk factors, the clinical manifestation of respiratory failure can be abrupt as is seen in our case.

Aquino et al. demonstrated that a reduction of more than 18 % in the cross-sectional area (CSA) of the upper trachea between inspiration and end-expiration has a Positive predictive value of 89–100 % for the diagnosis of trachobranchiomalacia. However, we did not perform an inhalation and exhalation CT scan to evaluate the change of the CSA of the airway to support our theory, because this will not change our management plan and patient comfort is our priority [12]. However, absence of other clinical findings of infection, airway reversible obstructions, pulmonary embolism, pulmonary edema and responsive to Heliox treatment. Airway narrowing is likely the main cause of this respiratory failure.

The respiratory failure in Morquio syndrome is a progressive development with multifactorial causes and irreversible pathology. Respiratory system management are reviewed and summarized in Table 2.

While there are several reports regarding invasive airway management of Morquio A syndrome, the timing of the surgery and requirements of specialized centers in performing these high-risk surgeries prohibited many Morquio patients from these advanced cares. In our case, the surgical management was reviewed with ENT physicians in a tertiary university affiliated specialized centre previously and this admission. She was evaluated for surgery in her 20's. However, she had Mallampati 4, open mouth 2 cm, hyomental distance of 6 cm with very limited neck extension. Both nasal and oral intubation failed due to obstructions in the upper airway. Not surprisingly, this time anesthesia and ENT declined the surgery for tracheostomy or stenting due to difficulty intubation and surgery, not to mention trachea and thoracic outlet reconstruction. Surgical management of the airway also pertains high risk of anesthetic and surgical failure. Stenting pertains high risk of re-obstruction and symptoms including severe coughing due to the high location of the stenosis [8,9,20].

In our case, BiPAP was the only way to support her life at a price of significantly decreased quality of life: Nasal pressure wound, abdominal distention and pain, limited room of ambulation, etc. Thus, with literature search and multidisciplinary discussion, three main pathologies were implicated in this case: narrowing of the airway as is shown in Fig. 1, OSA and restrictive thoracic wall based on the physical examination and history of Morquio syndrome. The VBG and response to BiPAP are also proofs of our theory.

These patients are always on the verge of respiratory disequilibria, mild disturbance may break the balance. Due to the acute onset of respiratory insult and failure, she became fully respiratory dependent on BiPAP with multiple complication associated with it. The complications of BiPAP created a vicious cycle of BiPAP dependence where BiPAP causes abdomen distention, lung compression and exacerbates respiratory failure that relies on BiPAP. See Fig. 2. She also complained of abdominal distention and pain, for which she required Hydromorphone and potential decreased respiratory drive. To facilitate O2 delivery through the significantly narrowed trachea and to break the vicious cycle of BiPAP dependence, we attempted Heliox to facilitate O2 delivery through the narrow airway

Table 2
Management of respiratory System Disorders in Morquio A Syndrome [28,30].

Non-invasive measures
Enzyme replacement therapy [31], haematopoietic stem cell transplantation [32], Gene therapy [5,33,34]
Vaccinations: pneumococcus, influenza, COVID-19 [35]
Early and aggressive treatment of respiratory tract infections [35]
Manual and mechanical techniques for cough assist [35]
Oxygen therapy including high flow oxygen therapy [35]
Heliox
CPAP/BiPAP [20,35]
Invasive measures
Early-stage tonsillectomy and adenoidectomy [36]
Intubation and mechanical ventilation [19,37–39]
Tracheal Stenting [8,9,20]
Tracheostomy [8,16,20,40]
Thoracic outlet organs reconstruction [9,10]

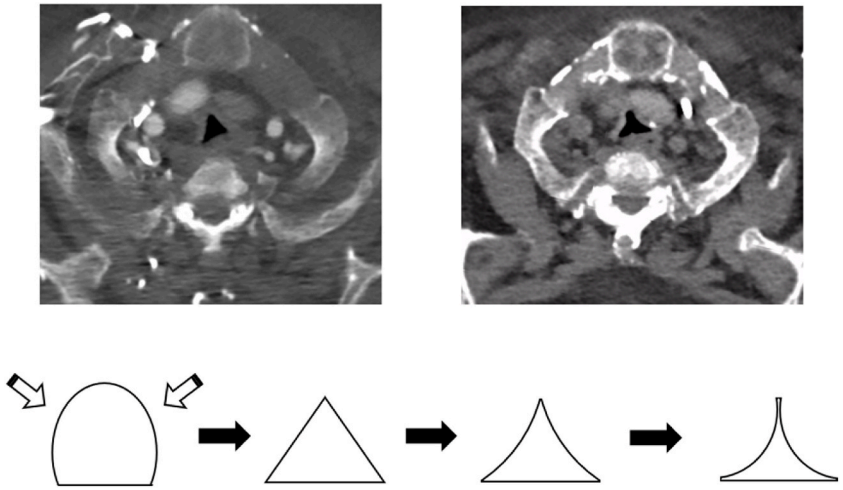

Fig. 1. Morphological changes of the trachea from 2013 (upper left) to 2023 (upper right). The size of the trachea at the level of thoracic outlet has no significant change, but the trachea in 2023 had a more significant morphological change due to either compression by the adjacent thyroid glands or innate tracheomalacia [8]. Scheme of morphological changes of the trachea over time. (lower).

and have a BiPAP break.

The limitation of the study is the lack of further treatment with Heliox due to the lack of supply in our centre. Whether prolonged use of Heliox would help her further reduce the time on NIV is unknown.

To the best of our knowledge, this is the first report of Morquio syndrome treated with Heliox, which lowers the density of the gas delivering to the lungs. It helps reduce the airway resistance makes the oxygen delivery through the narrowed trachea much easier according to the Poiseuille's Law: $Q = \pi Pr4/8\eta l$ (Q = flow rate, P = pressure, r = radius, η = viscosity, l = length) [41,42]. In our case, it helped the patient to wean off the BiPap for 4 hours per day. When she could have a break from the abdominal pain, pressure pain from the mask and less pain medication. Our study provides another option for airway narrowing in Morquio Syndrome. It improved the quality of life in our patient.

4.1. Patient perspective

4.1.1. Author Minghan Shi interviewed the patient and wrote this section based on the interview

I received treatment in the United States from a specialist for dwarfism. I had surgery on my neck and back. When I was 7–8 years

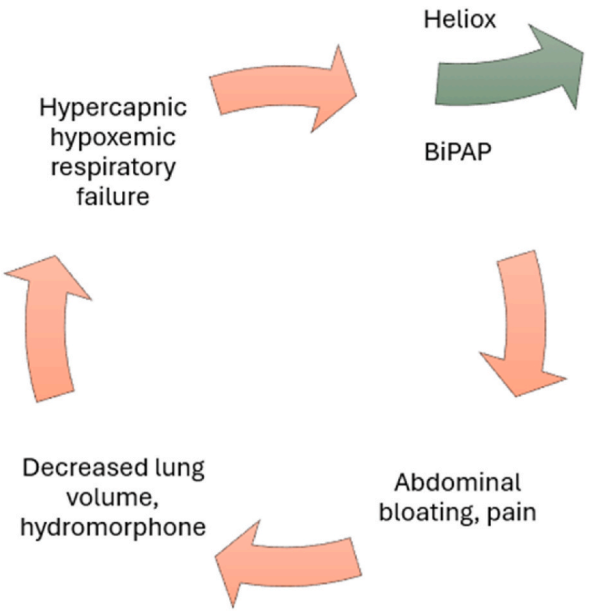

Fig. 2. Heliox played a role in breaking the vicious cycle of BiPAP dependence.

old, I needed scooters to help me walk. Later, at 14, I used a walker, and seven years ago, I switched to a power chair. I adjusted well to these changes. I also had frequent urinary tract infections, but they didn't greatly affect my life or work.

I did a lot of volunteer work. About ten years ago, I raised money for the Easter Seals by going down 14 stories in my wheelchair. I did this for four years. I also dressed up as Jingle's elf during Christmas, and volunteered as Flutter Bee and as a clown. I worked at the college information desk as well.

I was doing well with using BiPAP at night until last November, but things got worse after I returned from Toronto. I had to use BiPAP all day and had issues with my belly swelling, pain, and constipation. Using Heliox gas made me easier to breathe, and after a few days, I could go without BiPAP for a few hours. However, we ran out of Heliox, and I wished the respiratory therapist could see me more often than three times a week.

Recently, I was in the ICU due to difficulty breathing. Since being transferred from the ICU in February, I'm back to using BiPAP all day. I can only tolerate using Airvo for a few minutes. I hope I can receive more treatment with Heliox, but I was told that we don't have much stock in this hospital.

4.1.2. Learning point

Heliox provides a non-invasive option for BiPAP dependent Morquio A Syndrome patients due to trachea narrowing.

CRediT authorship contribution statement

Minghan Shi: Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Writing – original draft, Writing – review & editing. **Birubi Biman:** Supervision, Funding acquisition, Writing – review & editing, Conceptualization.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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