INFANTILE HYPOPHOSPHATASIA ASSOCIATED WITH RESPIRATORY INSUFFICIENCY.

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Introduction: We report the successful use of chronic mechanical ventilation (MV) in an infant diagnosed with infantile hypophosphatasia (IHPP) who developed chronic respiratory failure. This case illustrates the patient's presentation, course, and feasibility of use of chronic MV in patients with this condition. **Case Summary:** Patient is a 3.8 y/o biracial female with family history of adult form of HPP. She was born full term and presented at age 2 months (mo) with poor feeding, hypercalcemia, hyponatremia, and microscopic hematuria. The patient received nutritional supplements, calcitonin and was discharged home. She was diagnosed with IHPP at age 3 mo, and one month later G-tube/Nissen were placed due to failure to thrive. She was hospitalized 5 times during the first 5 mo of life for non-respiratory problems. She was re-admitted at age 6 mo for respiratory and urinary tract infection. She developed respiratory insufficiency, and was started on heated high flow nasal cannula (4LPM/1.0 F_IO_2). She began having frequent episodes of respiratory distress requiring intubation and MV. Initial ventilator settings were PRVC, rate-25, Vt-40 ml, PEEP-7, and F_IO_2-.35. She was tracheostomized at age 8 mo after 2 failed extubations. She was transitioned to LTV at age 15 mo after 5 unsuccessful attempts. She required placement back on Servo 300 at age 19 mo when she acquired RSV lower respiratory tract infection. She transitioned back to the LTV at age 20 mo and was discharged home 2 weeks later on ventilator settings of SIMV-PC/PS, rate-35, PC-18, PS-14, PEEP-6, and O_2-1.5 LPM. She was readmitted one mo later with respiratory infection and acute respiratory failure and changed to the Servo-i. She was treated with antibiotics and transitioned back to the LTV at age 37 mo and discharged 2 weeks later. **Discussion:** IHPP is a rare inherited disorder (1/100,000 births) characterized by defective bone mineralization. IHPP is diagnosed before age 1 year with onset of symptoms usually occurring within 6 mo and has 50% mortality rate. Diagnosis consists of hypercalcemia, decreased alkaline phosphatase, metaphyseal flaring, enlarged fontanelles, and wide cranial sutures. IHPP is considered fatal secondary to respiratory insufficiency or infection resulting from a defective thoracic bellows system as a result of demineralization of the ribs. This case illustrates that chronic respiratory failure in patients with this condition can be effectively treated with MV.

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HELIOX THERAPY IN THE TREATMENT OF MECHANICAL OBSTRUCTION SECONDARY TO CLOT FORMATION IN A PULMONARY HEMORRHAGE PATIENT.

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Introduction: A 3 year old, 15kg patient with a primary diagnosis of double outlet right ventricle with pulmonary stenosis, underwent a Kawashima procedure and hepatic vein baffle to the pulmonary circulation. The patient suffered a cardio-pulmonary arrest and was subsequently supported for five days with ECMO. After decannulation from ECMO, the patient suffered recurrent pulmonary hemorrhages leading to altered gas exchange and the need for high ventilatory pressures. Case Summary: Due to worsening respiratory status, a bronchoscopy was performed which revealed numerous blood clots in the airways likely causing a "ball-valve" effect. Approximately six hours following the bronchoscopy the patient's ventilatory support was escalated from PCSIMV 36/6 x 20 100% to PCSIMV 42/6 x 22 100% with 6mL/kg Vt. The patient continued to demonstrate a significant respiratory acidosis (ABG 6.82/241/77/37). Support was further escalated to 46/10, the flow graphics on the ventilator were suggestive of inadequate emptying of the lungs, therefore, the rate was reduced to 16 and heliox therapy was initiated at an 80/20 helium/oxygen mixture. Upon initiation of heliox the CO2 elimination rapidly increased from 54ml/min to 162ml/min and the Vt increased to 15ml/kg. Copious amounts of thick clot were suctioned from the endotracheal tube within minutes of heliox initiation, whereas prior to heliox, minimal clot was suctioned. Within four hours the patient's ABG improved to 7.30/63/41/30 and the ventilator was weaned to 38/8 x 16 with Vt 13ml/kg. Eight hours following heliox administration the ABG was 7.36/56/47/29 and the ventilator was weaned to 36/8 x 16 with a 70/30 heliox mixture. Heliox was discontinued thirty-six hours later and the patient was successfully extubated. Discussion: Heliox therapy has been used for treatment of severe bronchoconstriction and other etiologies that cause the airways to be narrowed. Our case exemplifies the use of heliox to improve gas exchange in a patient with blood clot causing partial obstruction of small airways. Heliox allowed us to improve gas exchange by improving laminar air flow, by-passing clot and re-inflating collapsed areas of lung. Additionally, heliox appeared to help facilitate migration of clot to the upper airways for removal, possibly due to increased peak expiratory flow rates.

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USE OF AIRWAY PRESSURE RELEASE VENTILATION WITH A TRAUMATIC BRAIN INJURED PATIENT.

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INTRODUCTION: Clinicians may be reluctant to use Airway Pressure Release Ventilation (APRV) when managing patients with traumatic brain injuries. There is a question that with the application of APRV, the PaCO2 cannot be successfully managed. Therefore, with an increase in PaCO2, a concomitant increase in intracranial pressures will occur. In addition, there is concern of increased intracranial pressure related to the increase in mean airway pressure with APRV. CASE SUMMARY: This case reviews a 62 year old male that was found down (presumed fall). On admission, he presented with a subdural hemorrhage and a Glasgow Coma Scale (GCS) of 7. An intraventricular catheter (IVC) was placed to monitor intracranial pressure (ICP) and drain cerebral spinal fluid. Due to a refractory increase in ICP, the patient required a craniectomy. The ventilator support was set to maintain PaCO2 between 35 and 40 cm H2O. The patient was on a Drager Evita XL with settings of SIMV/Autoflow/PS with a set respiratory rate of 25, tidal volume 550, PEEP 14, and FIO2 of 55%. On these vent settings, the patient's ICP ranged between 8-14 mmHg. The patient was then transitioned to APRV with settings of: Phigh 26, Plow 0, Thigh 4.2, Plow 0.55 (set respiratory rate of 13), FIO2 55%. Due to the improved alveolar ventilation that APRV provides, there was no need to match the SIMV minute volume (which was 13.8 L/min vs. 11.8). Analgesia and sedation remained the same with Fentanyl and precedex. In addition, hemodynamics remained stable after transition to APRV. DISCUSSION: Transitioning this patient to APRV demonstrated an improvement in oxygenation, ventilation and peak airway pressures with no untoward result of increased ICP or PaCO2. APRV has been useful as a lung recruitment mode, however, further research is needed to show the effect of APRV with TBI patients and ICPs and CO2 management.

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VITAL SIGNS	SIMV	AFTER APRV
ICP(mmHG)	7-11	4-12
CPP	74-92	74-89
ABG	7.45/35/150/26/2.7/98%	7.49/32/175/24/1.4/98%
MODE	SIMV 25/550/14	APRV 26/0 4.2/0.55
ATC	OETT: 8.0mm @100 compensation	OETT: 8.0mm @100 compensation
FIO2	55%	45%
MAP(cm H20)	21	24
SET RR (b/min)	25	13
TOT RR (b/min)	25	30
PEAK PRESSURE (cm H2O)	34-36	27
ETCO2	31	32
SPONT MV	0	12.4

1133202

HELIOX VIA OXYMASK FOR ADULT AIRWAY COMPRESSION.

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Introduction: Patients with lung cancer often experience dyspnea secondary to narrowing of the airway due to tumor growth. Heliox use is one method of reducing turbulence through a narrowed airway and relieving dyspnea. Case Summary: A 63 year old female, with progressive small cell lung cancer and persistent dyspnea was admitted to the ICU due to difficulty breathing with stridor and use of accessory muscles: RR 28, SaO2 94% on 2 lpm. The patient had a large right sided mass with bronchovascular encasement causing severe tracheal, bronchial, and vascular compression. There was a discussion of tracheostomy; however, upon evaluation it was evident the obstruction was too low for a tracheostomy to provide any relief. Within 12 hours of admission the patient was placed on an 80/20 mix of heliox via NRB mask at 10 lpm. The patient's dyspnea was improved, stridor cleared, and use of accessory muscles lessened; RR 20 SaO2 94% on 10 lpm HeO2 NRB. The patient was receiving daily radiation therapy treatments to reduce the size of the tumor. The challenges we faced were: high usage, limited availability of heliox and cost; $120.00 per tank, three tanks per day. Our usage rate would deplete the vendor's supply within two days and additional tanks were not available through any local vendor for 5 business days. In order to conserve gas, we attempted to reduce the heliox flow. The titration began with reducing the flow to the NRB mask to 6 lpm but the patient stated it wasn't enough, we increased to 8 lpm and she stated, "it would be tough" but she thought it might work, although she felt like it was a lot warmer. She was then placed on the OxyMask (Southmedic) with a flow rate of 4 lpm. Due to the design of the mask, the patient felt more comfortable than on the NRB mask at 8 lpm; RR 16 to 20, SaO2 96% on 4 lpm HeO2 OxyMask. We were able to maintain the flow at 4 lpm to 5 lpm for the duration of her stay; in addition we were able to place the patient on a nasal cannula at night while she slept. Thirteen days later the patient was discharged home to hospice care. Discussion: By using the OxyMask we were successfully able to titrate the heliox to achieve a therapeutic level that was comfortable and safe for the patient at half the flow rate effectively doubling our supply and achieving a 50% cost savings. We also have increased the supply of heliox in house and the vendor has increased their inventory in order to be able to care for similar patients in the future.

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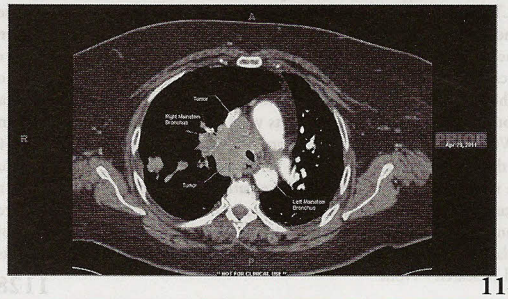

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