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Intensive Care Medicine

Correspondence

Inborn errors of metabolism (IEM) revealed by Reye's syndrome (RS)

Dear Sir,

Reye's syndrome (RS) [1] is an acute, rare but serious childhood encephalopathy in which vomiting and coma develop 3–5 days after a viral prodromal disease. Selective hepatic dysfunction is a constant finding. Panlobular microvesicular fatty changes and mitochondrial alterations are characteristic of the disease. Many aetiologic factors have been implicated (viruses, drugs, toxic substances). Recent reports have stressed the importance of early screening for IEM masquerading as RS [2–5]. This justifies a systematic metabolic evaluation on admission, performed in parallel with other investigations. In our unit, this has revealed two fatty acid oxidation defects and 1 partial ornithine transcarbamylase (OTC) deficiency out of 4 consecutive cases of RS observed during a 4-year period (2000 admissions):

A 3 1/2-year-old girl presented with stage-3 coma and seizures during the course of a common cold. Previous history included muscle weakness and hypoglycemia. Laboratory studies showed hypoketotic hypoglycemia. ASAT 160 IU/l, ALAT 100 IU/l, normal prothrombin time, hyperammonemia (145 mol/l), low serum carnitine levels (free 4 µmol/l, total 25 µmol/l, F/T ratio 16%), and medium chain dicarboxylic aciduria (C6, C8, C10). Liver biopsy showed macrovesicular perilobular and microvesicular centro-lobular steatosis. Fibroblast oxidation of labeled C-14 was 39% of normal with C8, normal with electron transfer flavoprotein, suggesting an electron transport defect. Outcome was satisfactory.

An 11-year old boy developed a cold, vomiting and stage-4 coma. Laboratory data showed: pH 7.5, PCO_2 31 mmHg, HCO_3^- 20, ammonemia 390 mol/l, ASAT 48 IU/l, ALAT 78 IU/l, prothrombin time 39% and oroticuria 1836 mg/g creatinine. Liver biopsy revealed micro- and macro-vesicular steatosis with moderate glycogenic depletion without hepatocellular necrosis or mitochondrial changes. The patient died 5 days later but residual liver OTC activity was not studied. Partial OTC deficiency of late onset was confirmed by protein challenge of the sister and mother (Basal oroticuria: 18/22 µg/mg creatinine respectively (normal <2.9)); 6 h post-challenge: 83/87.

A 13-month-old boy was admitted with intense vomiting and coma following a viral infection. Laboratory studies showed: ASAT 27500 IU/l, ALAT 12000 IU/l, prothrombin time 10%, ammonemia 230 mol/l and hypoketotic hypoglycemia. Liver biopsy was normal. He recovered completely but relapsed two years later. Serum carnitine/amino acids and urine GC/MS were unremarkable. Ketone bodies increased normally after a medium-chain triglyceride callenge, but not after long-chain trigylcerides. An unidentified defect in long chain fatty acid oxidation is suspected (normal hepatic carnitine palmitoyl transferase activity).

IEM must be searched in any case of RS in order to exclude mainly β-oxidation defects, urea cycle defects and organic acidemias [2–5]. Since clinical findings indicative of an underlying IEM [3] are non-specific, metabolic studies should be done for *all* patients with RS. Serum and urine samples should be taken *at presentation* and specific treatment administered immediately. Simple lab tests can be of great value [3]: hypoketotic hypoglycemia indicates possible fatty acid β-oxydation enzyme defects; whereas alcalosis, severe hyperammonemia contrasting with mild hepatic cytolysis, a urea-cycle defect. Furthermore, macrovesicular fatty changes without ultrastructural mitochondrial changes should araise suspicion of an IEM.

Yours faithfully,

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Continuous flow modification for Siemens Servo 900C ventilator

Dear Sirs,

I read with interest the correspondence entitled, "CPAP with a Siemens Servo 900 C Ventilator during weaning in infants" [1].

I would like to report on our modification of the Siemens Servo 900 C Ventilator, which we have used successfully since 1986 on infants and children up to 16 kg, both in the pressure control and volume delivery modes. These patients had varying conditions which included: cardiovascular surgery, chronic lung disease, neuromuscular disorders and difficulty in weaning individuals. Our goal(s) for this type of application were: (a) To limit the work of breathing through use of non-demand valve effect [2–5]; (b) Prevention of dysynchrony; (c) Maintenance of PEEP, with the presence of a significant leak around an artificial airway; (d) To enhance our ability to wean ventilator dependant patients.

We support the finding that gas flow meets inspiratory demands, and that no negative effort is necessary to initiate flow, as well as the findings with the monitoring and alarm functions.

Please note on our schematics, we have included an anti-asphyxia valve, Fig. 1. Should inadvertent loss of gas flow occur, the Siemens Servo 900C does not contain an internal anti-asphyxia valve. We do realize the Servo 900C contains an internal loss of gas supply alarm, yet the anti-asphyxia valve is still utilized should that occur. This valve is a variable orifice device calibrated at 5 cmH_2O. A negative pressure created

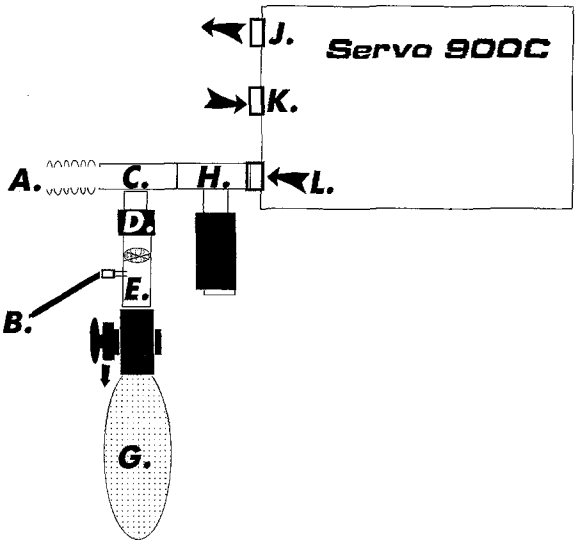

Fig. 1. Schematic diagram of Siemens Servo 900C. Continuous flow modification. *A* Ventilator inspiratory tubing; *B* continuous gas flow connection; *C* airlife disposable Trach "T" piece; *D* rubber adaptor; *E* airlife straight connector with one way valve; *F* Mapleson flow adjustment valve; *G* one liter anesthesia reservoir bag; *H* airlife disposable trach "T" piece; *I* Boehringer 5 cmH_2O anti-asphyxia valve; *J* expiratory gas outlet; *K* expiratory gas flow from patient; *L* inspiratory gas flow to patient. *Product Sources:* Siemens Servo 900C Ventilator, Siemens Medical Systems, Inc., Schaumberg, Illinois. Ventilator Inspiratory Gas Tubing, Disposable Trach "T" piece, Straight Connector With One-Way Valve, (*A. C. E. H.*) Baxter Healthcare, Valencia, California. Continuous Gas Flow Connection (*B*), Hudson-RCl, Temecula, California. 15 m Rubber Adaptor (*D*), Intec Medical, Inc., Blue Springs, Missouri. Anesthesia Reservoir Bag-Mapleson Flow Adjustment Valve (*F, G*), Vital Signs, Inc., Totowa, NJ. Boehringer Anti-Asphyxia Valve (*I*) Boehringer Laboratories, Norristown, PA

in the circuit greater than -5 cmH_2O, opens the orifice and allows for entrainment of room air into the patients circuit. The addition of this valve requires the ventilators sensitivity be set at a level less than -5 cmH_2O, to a point where the patient no longer triggers the ventilator. This prevents inadvertant opening of the valve as well as serving as a back-up gas flow source.

Also included in our modification is a Mapleson Scavenger Valve, which allows for maintenance of PEEP or CPAP while having a constant blended flow of gas supplying a one l reservoir bag (Fig. 1). To operate this, fresh gas flow should be turned on at 10–15 l/min, with the scavenger valve completely open. The scavenger valve should then be closed until the one-way valve remains open and if in volume or control mode, it should remain open between the ventilator positive pressure breaths. Upon delivery of a positive pressure breath, the valve should close *completely.* This valve should eliminate the need for a 4 l bag. This entire set-up is prehumidifier, and the gas flow serving the bag should come from the flowmeter off the ventilator blender outlet, this allows for equal FiO_2 on both spontaneous and mechanical breaths.

When employing this modification, the PEEP control adjust may have to be fine tuned. The constant flow results in inadvertent PEEP which requires readjustment of the PEEP control knob. PEEP control may be enhanced with your scavenger valve while being read off the analog. It may be impossible to have PEEP of O due to constant flow. The analog deflection should also be utilized to test adequacy of gas flow upon patient inspiration.

Deflections greater than -1 to -2 cmH_2O, may require an increase in flow. This does require ongoing assessment as patient sleep states, etc. vary.

As you described, this modification does mimic the characteristics of a constant flow pediatric or neonatal ventilator. As stated before, in our experiences, it has been used on a range of patients without incidence. It has also allowed us another option in our efforts to mechanically ventilate the most difficult infants and children.

I would like to thank Mr. Joe Lore, BS, RRT who, in 1986, was Education Clinical Coordinator at Children's Hospital of Philadelphia and advised us upon initiation of this modification.

Yours faithfully,

J. L. Swegarden

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Unilateral pulmonary damage

Dear Sir,

unilateral acute lung unjury (UALI) constitutes an infrequent clinical manifestation of acute parenchymal respiratory failure. Such pathologies are characterized by the separation of the two lungs in hemi-systems with different functional and anatomical characteristics [1].

In this pathophysiological context, Independent Lung Ventilation (ILV) is an indicated ventilatory mode [2–4].

Recently a 16-year-old-female, with closed chest trauma, was admitted to the Emergency Department of Torrette Regional Hospital in Ancona after a road accident. A chest X-ray film showed bilateral diffuse interstitial involvement due to pulmonary edema with an alveolar aspect in the supra-basal and middle pulmonary field of the right lung and with the presence of a mild Pneumothorax.

The initial treatment consisted in progressive increases of the FiO_2 by a Ventimask, and in SaO_2 monitoring by pulse-oximetry. After an initial improvement, the SaO_2 decreased progressively. CPAP was started using a face mask with 10 cm H_2O pressure. Even this therapeutic procedure failed, and tracheal intubation became necessary. A bronchoscopy showed bloody secretions coming from right superior and middle lobar bronchi.

In view of the cardiorespiratory deterioration during IPPV and CPPV a decision was made to use ILV. The following ventilatory pattern used was: IPPV on the left side, with correction of tidal volume and respiratory rate according to blood gas analysis (BGA), and 20 cm H_2O PEEP on the right side, without mandatory breaths.

Initially FiO_2 0.6 was applied to both lungs; it was reduced to 0.4 when the oxygenation was stabilized.

This treatment was maintained for 15 h; then, a spontaneous ventilatory pattern was established, using 20 cm H_2O CPAP and a FiO_2 0.4 for both lungs, because the X-ray film showed a complete resolution of the unilateral lung damage.

During weaning, the PEEP was progressively reduced, by steps of 5 cm H_2O/h. The patient was extubated 24 h after the trauma. After 15 days, X-ray, CT-scan and pulmonary function tests showed a complete recovery.

ILV can be performed in many ways, according to clinical conditions of the two lungs [5]. The originality of this case is due to the use of artificial ventilation by IPPV in the healthy lung only, while a 20 cm H_2O