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The successful use of continuous negative extrathoracic pressure in a child with Glenn shunt and respiratory failure

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Abstract Following a Glenn shunt, an infant required mechanical ventilation (IPPV) for pneumonia, a phrenic nerve palsy and chylothoraces. In order to improve her deteriorating clinical condition, we used continuous negative extrathoracic pressure (CNEP) to minimise the deleterious effects of IPPV on pulmonary blood flow. She was sucessfully weaned from IPPV and supported with CNEP.

Key words Ventilation, negative extrathoracic pressure · Cardiac surgery · Glenn shunt · Pulmonary infection · Respiratory syncytial virus · *Pseudomonas aeruginosa*

Introduction

Intermittent positive pressure ventilation (IPPV) and positive end-expiratory pressure (PEEP) are frequently required as part of the management of respiratory failure. In patients without a functioning right ventricle, pulmonary blood flow is passive and small amounts of PEEP have deleterious effects on the cardiac output [1]. We describe a patient with both these conditions whose management was extended to include continuous negative extrathoracic pressure (CNEP).

Case report

A female term 3.7 kg infant was found to have pulmonary atresia and an intact ventricular septum. In the neonatal period, she underwent an atrial balloon septostomy followed by a 4-mm modified left Blalock-Taussig shunt (BTS) and open pulmonary valvotomy. She could not be weaned from mechanical ventilation, and 12 days after surgery angiography revealed kinking of the distal end of the left modified BTS. A right 4-mm modified BTS was created and she was extubated after 24 h.

At 12 weeks of age, her atrial septal defect had become severely restrictive and she underwent further palliative surgery. The fossa ovalis was excised, the right BTS was ligated and divided, and a modified bidirectional Glenn procedure (superior vena cava-right pulmonary artery anastomosis with detachment of the SVC from the right atrium) was performed. Her postoperative course was complicated by recurrent right chylothorax and palsy of the right phrenic nerve with paradoxical movement (confirmed by subsequent ultrasound investigation and measurements of phrenic nerve conduction). The right hemidiaphragm was plicated, after which she was extubated uneventfully. The chest drainage of chyle stopped once she was fed on medium-chain triglyceride milk.

She was discharged at 16 weeks of age (4.5 kg), only to present 2 days later with poor feeding, dyspnoea, irritability, swelling of the head and neck and an oxygen requirement (peripheral oxygen saturation 35–60%). Cardiac catheterisation showed no obstruction of SVC blood flow. Chest X-ray showed complete opacification of the right hemithorax with minimal shift of the mediastinum. The right chest was aspirated but failed to drain fluid. After 2 days she was intubated and ventilated for a CT scan, which showed pleural thickening and consolidation and collapse of the right lung. Bronchoscopy failed to remove thick secretions. Sedation was adjusted to the point where she tolerated the endotracheal tube and would permit endotracheal but spontaneous ventilatory effort was conserved. PEEP was kept at 1–2 cm H_2O. Oxygenation did not improve; PaO_2 ranged from 3.0 to 4.5 kPa in 50% O_2, and the $PaCO_2$ rose progressively (Fig. 1). *Pseudomonas aeruginosa* was grown in

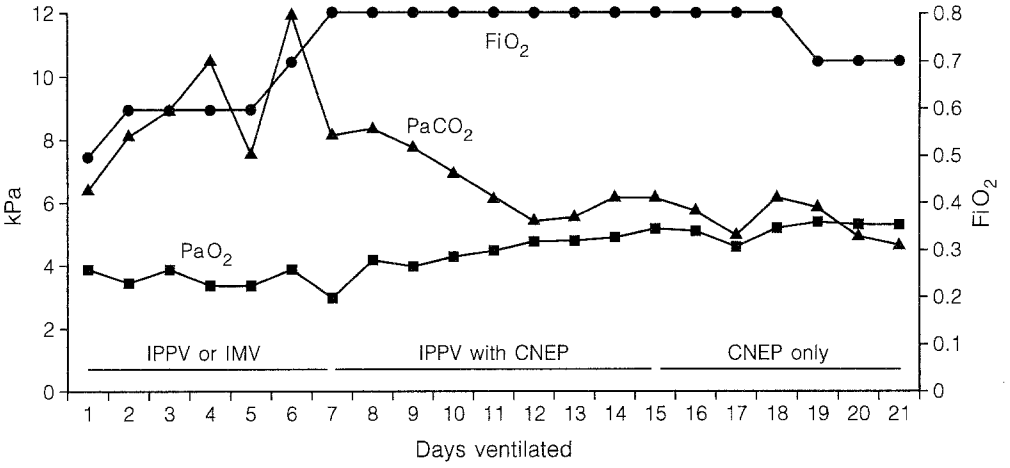

Fig. 1 Arterial blood gas analysis. The changes in pCO_2, pO_2 and FiO_2 during the period of respiratory support

sputum culture and respiratory syncytial virus was identified by immunofluorescence. Netilmicin and azlocillin were started. Partial reexpansion of her right lung after 2 days was not associated with any improvement in PaO_2. In order to reduce mean intrathoracic pressure attempts were made, albeit unsuccessfully, to synchronise intermittent mandatory ventilation with the patient's airflow-triggered positive pressure breaths using an SLE 2000 time-cycled pressure-limited ventilator (SLE, South Croyden, Surrey, UK). Increases in alveolar ventilation with full IPPV served only to worsen oxygenation. Her respiratory acidosis was fully compensated (pH 7.39 −7.42, HCO_3^- 41.7−47.7, base excess +16.3−+20.3).

Ventilation and physiotherapy had achieved reexpansion of her right lung, but this was not associated with any improvement in gas exchange. We postulated that IPPV was impeding pulmonary blood flow via the Glenn shunt and that her cerebral irritability was due, in part, to cranial venous engorgement, exacerbated by IPPV and systemic hypoxaemia. This combination of problems suggested that negative extrathoracic pressure ventilation [2] might allow alveolar ventilation to take place at lower positive intrathoracic pressures while maintaining the distending gradient across the alveolar bed needed to maintain the process of lung expansion. Additionally, at lower peak ventilator pressures a larger part of the ventilator cycle would then take place at a negative extrathoracic pressure, thus improving systemic venous return to the lungs, especially from the head, where the venous drainage depended on the state of the SVC run-off into the lungs.

IPPV, initially at 22/2 cmH_2O, rate of 35 and I:E ratio of 1:1.3, was continued and continuous negative extrathoracic pressure (CNEP) according to the method of Samuels and Southall [2] started at −6.0 cmH_2O. Simultaneous with the onset of CNEP, peak inspiratory pressure (PIP) was reduced by 6 cmH_2O and the PEEP, reduced to zero. Immediately her agitation disappeared and she settled into a light sleep. Within 4 h, her spontaneous ventilation rate had fallen from 42−60/min to 25−30/min, without any increase in $PaCO_2$, and by the following day to 9−20/min without any change in the ventilator cycle. Other signs of respiratory distress improved: arterial oxygen saturation (SaO_2) rose from 59−64% to more than 70% and sedation was no longer necessary. The pH rose minimally to between 7.44 and 7.48, and the base excess fell to normal over 5 days. The progress of other measured variables is displayed in Fig. 1. Upper torso and facial swelling resolved over 24 h. Transient right upper- and middle-lobe collapses were treated with conventional manual ventilation and physiotherapy. Eight days after commencing CNEP the infant was extubated but remained in negative pressure of −10 cmH_2O for a further 5 days before weaning of CNEP commenced. This was achieved by reducing CNEP by 1 cmH_2O per day. After reaching 4 cmH_2O she was allowed out of CNEP for 1 h, this being extended by 1 h per day until she was in CNEP at night only. To familiarise local staff with her apparatus, she was discharged to her base hospital, from which she was subsequently sent home at 7 months of age with her parents being fully versed in her management. Over the course of the next 5 months, whilst phrenic nerve recovery continued, whe was treated by her parents [3]. At home CNEP was applied at −6 cmH_2O at night, supported by a programme of transcutaneous PO_2 monitoring [4]. Two respiratory tract infections during this time responded to antibiotic therapy. At 12 months of age, phrenic nerve conduction studies were almost normal and overnight recordings of oxygenation and transcutaneous pCO_2 showed normal, stable values. CNEP was therefore successfully discontinued.

Discussion

Following a Glenn procedure, blood flow through the lungs is influenced by the pressure gradient from SVC to the left atrium and the pulmonary vascular resistance (PVR). Respiratory failure, consequent upon pneumonia and lung collapse, aggravated by a stiffened pleura and phrenic nerve palsy, was initially managed with IPPV only, but this resulted in worsening of oxygenation and oedema of the head. Efforts aimed at reducing these adverse effects of IPPV by reducing mean positive intrathoracic pressure failed, as evidenced by the rising $PaCO_2$ and persistently low PaO_2, indicating inadequate alveolar ventilation despite reexpansion of her right lung. This suggested that IPPV had worsened ventilation perfusion (V/Q) matching, probably on the perfusion side of the ratio.

Negative extrathoracic pressure support in the spontaneously breathing patient may produce lung distension (compensating for paradoxical diaphragmatic movements in those with phrenic nerve palsies) without increasing pulmonary vascular resistance [5], and can improve effective pulmonary blood flow [2, 6]. Other beneficial effects of CNEP include an improved residual volume, an in-

crease in vascular cross section contributing to a reduction in PVR and, by compensating for the normal infants' compliant chest wall, reduction in the work of breathing and lessening of diaphragmatic fatigue. By reducing intrapleural pressure, CNEP will ameliorate venous return.

In our patient CNEP was associated with a reduction in the signs of respiratory distress, which we speculate was coincident with a reduction in the work of breathing. The $PaCO_2$ fell, indicating an improvement in alveolar ventilation, and the PaO_2 rose, indicating an improvement in V/Q matching. The resolution in upper-body oedema presumably followed a reduction in PVR and increased SVC drainage.

Changes in pulmonary blood flow in time with the respiratory cycle have recently been reported in patients with a total cavopulmonary shunt. During spontaneous ventilation, Redington et al. [7] confirmed the augmentation of forward flow with inspiration and its reversal during a Valsalva manoeuver. Penny et al. [6], using Doppler echocardiography to measure pulmonary artery blood flow, showed that this increased with the application of negative extrathoracic pressure and reversed when extrathoracic pressure became positive.

The eventual natural recovery in phrenic nerve conduction seen in our patient has been described previously in children supported with negative pressure ventilation [8].

In conclusion, we would suggest that CNEP with IPPV or CNEP alone may provide a useful form of respiratory support for those patients in whom PVR needs to be kept at a minimum and pulmonary blood flow enhanced.

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