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CASE STUDY

Walking with continuous positive airway pressure

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ABSTRACT: A ventilator-dependent child had been in the paediatric intensive care unit (PICU) ever since birth. As a result, she had fallen behind considerably in her development.

After 18 months, continuous positive airway pressure was successfully administered *via* a tracheostomy tube with a novel lightweight device. This enabled her to walk in the PICU. With this device, the child was discharged home where she could walk with an action range of 10 m. Subsequently, her psychomotor development improved remarkably.

To the authors' knowledge, this is the first case report of a patient, adult or paediatric, who could actually walk with a sufficient radius of action while receiving long-term respiratory support.

KEYWORDS: Boussignac, chronic lung disease of infancy, continuous positive airway pressure, tracheobronchomalacia, tracheostomy

The current study presents a case of a child with chronic lung disease (CLD) of infancy and tracheobronchomalacia (TBM). TBM with or without CLD is associated with prolonged ventilator dependency, even when only continuous positive airway pressure (CPAP) is needed [1, 2]. Usually a general-purpose ventilator, a dedicated noninvasive ventilator or a CPAP machine applies CPAP. Regardless of whether CPAP is applied *via* a full-face mask, nasal mask, nasal prongs or a tracheostomy tube, patients who are otherwise awake and mobile will be bound to their bed or wheelchair. In children especially, this immobilisation will impede the development of motor skills as well as many other skills.

Recently, a CPAP device (Boussignac system; Vygon, Ecouen, France) has become available that can be directly connected to the tracheostomy tube and is driven only by compressed air. The present study describes the successful application of the Boussignac CPAP, which enabled the patient to become mobilised and walk.

CASE REPORT

Pentalogy of Cantrell was diagnosed, in a female born at 36 weeks gestational age and with a birthweight of 2,000 g. This is a rare congenital malformation characterised by: 1) a lower sternum defect; 2) an anterior diaphragm defect; 3) a pericardium defect; 4) an omphalocele; and 5) cardiovascular abnormalities. She had been ventilator dependent since birth due to TBM and developed CLD due to a complicated abdominal closure with need for mechanical ventilation at high pressures. At the age of 4 months, a tracheostomy was performed because prolonged ventilatory support and weaning was expected. In the months following the tracheostomy, weaning attempts failed repeatedly due to recurrent hypoxaemia and hypercapnia. Assessment at the age of 13 months showed a psychomotor development compatible with the age of 6 months.

During weaning with conventional CPAP apparatus, the heavy tubing severely restricted the patient's mobility. For that reason, Boussignac CPAP (fig. 1) was introduced at the age of 18 months to stimulate psychomotor development. Pressurised air and oxygen with a final oxygen fraction of 30% and a flow of 15 $L \cdot min^{-1}$ was used to deliver 5 cmH_2O of CPAP to the tracheostomy tube. Humidification was supplied with an ordinary syringe infusion pump supplying 3 $mL \cdot h^{-1}$ of sterile water to the Boussignac CPAP device. Boussignac CPAP, combined with extended tubing, offered the child an action range of 10 m so she could play and develop motor skills. Over a period of 6 months, Boussignac CPAP was gradually extended to all hours that the patient was awake. During sleep,

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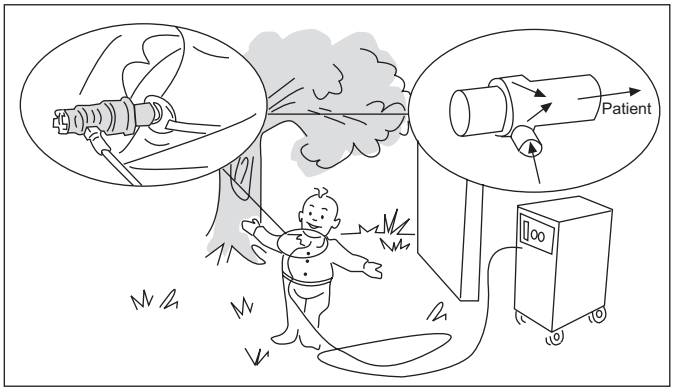

FIGURE 1. Schematic diagram showing the delivery of continuous positive airway pressure (CPAP) to an infant at home. The child learned to walk while on long-duration daytime CPAP support with a novel system. Pressurised air was generated by a transportable air compressor powered by electricity from the mains connection (height 0.85 m) and transported through thin tubing with a length of 10 m. CPAP (4 cmH_2O) was generated *in situ* by the Boussignac valve that was connected to the tracheal canula. Due to the geometry with which the compressed air is injected into the hollow tube (inner diameter 0.5 inches or 1.2 cm), a virtual ''valve'' is created.

the child was mechanically ventilated. Supplemental oxygen was gradually decreased to zero. In the same period, the child was fully mobilised with Boussignac CPAP and her psychomotor and mental development improved dramatically.

Finally, after an admission of 2 yrs at the current authors' paediatric intensive care unit, the child was discharged home. She was still receiving Boussignac CPAP during the day and pressure-controlled ventilation (18/4 cm of H_2O; frequency $25 \cdot min^{-1}$; inspiratory oxygen fraction 0.3) during the night. Pressurised air was generated at home by a medically certified compressor (Jun-air, Nørresundby, Denmark). At home, the time spent on Boussignac CPAP was gradually tapered over 1 yr. No technical problems were encountered, and especially no obstructions of the tracheostomy tube occurred. At the age of 2.5 yrs, new testing showed that the child's psychomotor development had returned to normal according to calendar age. At the age of 3 yrs, the child could breathe adequately *via* a tracheostomy tube without additional support while awake. At present (3.5 yrs), she is being weaned from ventilatory support during the night.

NOVEL CPAP DELIVERY SYSTEM

The Boussignac CPAP system is a Food and Drug Administration approved, simple and lightweight (10 g) disposable plastic device without moving parts. It was developed to deliver CPAP for face masks [3]. In addition to generating positive pressure, the system also allows the administration of high inspiratory oxygen fractions and humidification. Since CPAP is generated *in situ*, the system only needs compressed air or oxygen delivered through thin tubing that can be of any length. CPAP varies linearly when the flow is applied. This contrasts with the short and heavy tubing that characterises other CPAP systems. Although marketed for application to face masks, Boussignac CPAP can be easily applied to tracheostomy tubes as well. In adult patients who are weaned from respiratory support with a tracheostomy tube, Boussignac CPAP is already frequently used. The unique properties of the Boussignac system result in more freedom of movement for these patients. The low weight of the overall system also reduces mechanical forces on the trachea and surrounding structures. Since the Boussignac ''valve'' is only a virtual valve with no moving parts, it cannot be obstructed by sputum, a common problem in tracheostomy patients. Moreover, given the open design of the Boussignac valve, the patient can still breathe freely without any resistance if flow is interrupted.

The Boussignac CPAP system has one major disadvantage, which is the noise that is produced (55 dB measured at 1 m at a CPAP level of 5 cmH_2O). The noise is related to the flow that is applied.

DISCUSSION

With Boussignac CPAP, it was possible to allow a patient on long-term ventilatory support to walk with a sufficient radius of action. With the combination of TBM and CLD, CPAP was applied 24/24 as treatment for the TBM to splint the airways. Ventilation with additional oxygen during the night served as treatment for CLD. Children who are ventilator dependent due to CLD and/or TBM have a good chance to ultimately become independent of long-term ventilatory support, albeit after 1.6–2.3 yrs [4].

Thus, CLD and TBM are associated with a great impact on intensive care resources and healthcare utilisation [5]. Therefore, it is important to consider and develop alternative techniques that allow early mobilisation and discharge of these patients.

Although the hospital, and especially the intensive care unit, is an unsuitable environment for the developing child, discharging these patients is difficult [1, 4]. In the present study, a significant delay occurred before the patient could be discharged, in part as a result of the process of obtaining certification of the equipment for use at home.

With the Boussignac CPAP system, the child was successfully mobilised and discharged home. Although a direct relationship between mobilisation with the Boussignac CPAP and the improved psychomotor development cannot be proven, it is obvious that the freedom to move stimulates psychomotor development in children. Assisted ventilation during exercise has been applied in adult chronic obstructive pulmonary disease patients [6]. However, these studies have been carried out on a treadmill or bicycle ergometer. In these cases, the patient cannot be considered to be mobile. Although portable ventilators have been evaluated in adults [7], and might be used under parental guidance, such systems are too heavy to be used by a playing child. The possibility to mobilise CPAP-dependent patients may improve the quality of life of children and adults. The relative safety of the open design of the Boussignac system is reflected by the fact that no tube obstructions occurred over a period of 18 months.

Boussignac CPAP may be of use for the temporal support of adult patients with reversible respiratory insufficiency. ENGOREN *et al.* [8] described that out of 429 adult patients who received a tracheostomy in a tertiary care centre, as many as 143 were discharged while ventilator dependent. Although

the authors do not describe the percentage of patients who had reversible CPAP dependency, in the current authors' experience this number may be considerable.

In conclusion, the Boussignac system was successfully used to treat a continuous positive airway pressure-dependent patient with a tracheostomy. This allowed the patient to walk and achieve a normal level of development. Children especially may benefit from this therapy so they can develop in a more normal way.

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