Case Report

Long term intubation and successful weaning in two children with Guillain-Barre Syndrome

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ABSTRACT

No definite criteria exists in Guillian-Barre Syndrome in children regarding prolonged ventilation through an endotracheal tube without tracheostomy and successful weaning using a T-piece. Here we report two such cases of Guillian-Barre Syndrome requiring prolonged intubation for 56 days and ventilation for 30 days and ultimately successfully weaning them using the T-piece. Both the children eventually made a complete recovery, highlighting the point that in children prolonged intubation and ventilation using the portex tube is equally good, if not, better than tracheostomy with its attendant risks.

Keywords: Guillain-Barre Syndrome, prolonged ventilation, weaning, T-piece, extubation.

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Previous studies on difficulties in weaning ventilatory dependent pediatric patients are surprisingly rare. The data on severe respiratory insufficiency occurring in Guillain-Barre-Syndrome (GBS) leading to mechanical ventilation are scanty.[1-6] Immuno-modulation with infusions of intravenous gamma globulin (IgG) or plasma exchange treatment foreshorten the disease course in majority of the cases of pediatric GBS and do not require mechanical ventilation.[7] Since the incidence of pediatric GBS requiring assisted ventilation are small, prospective data on the indications for and the course of mechanical ventilation, particularly the weaning procedure is rare.[8,9] Also in most intensive care units, tracheostomy rather than prolonged intubation is the dictum. There are hardly any reports on prolonged intubation and successful weaning making use of T-piece, and increasing periods of pressure support ventilation (assist-control mode). We are reporting here two cases, which presented with respiratory failure following GBS requiring mechanical ventilation for prolonged periods. We were successful in weaning and extubating both the children using the T- piece.

Case Report.

Patient 1. A 2 year old female child was admitted in the pediatric intensive care unit of our hospital with one day history of progressive ascending symmetric weakness with loss of voice, difficulty in swallowing and head holding. Both upper and lower limbs were flacid and all the tendon reflexes were lost. There was also autonomic dysfunction. Respiration was mainly abdominal and chest expansion was less than 0.5 cms. The cerebrospinal fluid examination showed albumino-cytological dissociation (high protein with no cells). Nerve conduction study revealed grossly reduced motor and sensory nerve conduction with prolonged latencies. On these findings a diagnosis of GBS was made

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fulfilling the established clinical, biochemical and electro-physiological criteria.[10,11]

The patient received intravenous immunoglobulin (IVIg) 400 mg/kg for five days. However, two days after admission, the child went into respiratory failure and was subsequently ventilated. After three weeks, the child was showing features of recovery with good head and neck control and upper extremity movements. At 30 days of admission, the child was on pressure support mode with a peak inspiratory pressure (PIP) of 10 and FiO2 of 21%. On day 37 the child was extubated. After extubation she was using abdominal muscles to breathe and started to retain PCo2 necessitating reintubation and ventilation after 20 hours of extubation.

After unsuccessful extubation it was decided to try the weaning procedure using T-piece and decreasing periods of positive pressure ventilation. We designed simple clinical parameters to decide when to interrupt the periods of spontaneous breathing:- 1. Respiratory rate exceeding 60 per minute; 2. Intolerable breathlessness; 3. Hypotension or hypertension; 4. Blood gas pH below 7.29; 5. PaCo2 increase of more than 1 KPa. Minute volume was not considered as it will change with the respiratory rate and vital capacity was not measured due to lack of facilities. Spontaneous breathing was interrupted whenever one of the above-mentioned conditions existed.

The weaning procedure was started on day 51 of ventilation using T- piece. The period of spontaneous breathing gradually increased so that on day 56 of ventilation, the child was not requiring interruption of spontaneous breathing and was able to breathe 24 hours continuously without mechanical ventilation. During the ICU stay, chest physiotherapy was performed using standard techniques of manual vibration, postural drainage and bronchial suctioning of secretions as and when required. Prompt treatment of infections and good nutritional support were also given. The child was successfully extubated on day 56 of ventilation. She did not develop any complications of prolonged intubation and ventilation and had recovered from her primary illness in six months.

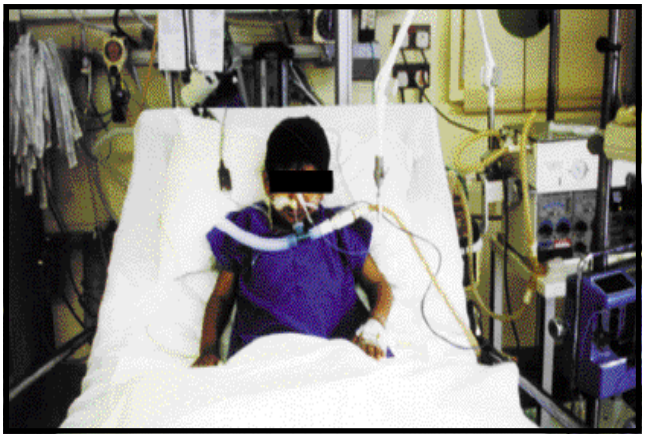

Figure 1 - Child with GBS being weaned from the ventilator. Note the corrugated reservoir tube connected to the expiratory limb.

Patient 2. A six and half year old boy was referred from a peripheral hospital with eight days history of body aches and pains, four days history of weakness and hypotonia, inability to walk, drooling of secretions, inability to talk and one day history of breathing difficulty, necessitating positive pressure ventilation. A diagnosis of GBS was made on the basis of progressive weakness of both upper and lower limbs, absence of fever at onset, areflexia, relative symmetry of signs, autonomic dysfunction and the cerebrospinal fluid showing albumino-cytological dissociation (Protein of 5gm/l with no cells). Nerve conduction study showed grossly prolonged latencies with markedly reduced compound motor action potential amplitudes in all motor nerves of the upper and lower limbs. No sensory action potentials were elicited.

The child received intravenous immunoglobulin and volume control ventilation with other supportive measures. Three weeks after admission, the child was showing features of recovery and at 24 days of ventilation weaning procedure was tried using T-piece, as in patient 1, using increasing periods of spontaneous breathing and was extubated successfully on day 30 of ventilation.

Discussion. The occurrence of respiratory failure is a life-threatening complication of GBS. Epidemiological surveys of GBS or selected case series in critical care literature reports about 10% (4-22%) of patients with GBS requiring ventilator support at some point in the course of their illness.[1,7] In most ICU's, it is customary to tracheostomize these children.[2] Complications of tracheostomy in children include injury to cartilage, vessels and pleura, fistula, hemorrhage, decannulation difficulties, tracheal granulations and later wound infection and scar.[2] In our unit, with the modern portex, non-cuffed endotracheal tubes, using advanced skills in intubation and fixation of the tube, ably supported by meticulous nursing care, we prefer endotracheal intubation rather than tracheostomy in infants and young children, even on prolonged ventilation.

In our first patient, the child was ventilated for 56 days and following extubation there was no hoarseness of voice, throat pain or difficulty in swallowing and the child had a smooth post-extubation course. No laryngeal injury or subglottic stenosis developed. Child has been followed up for more than 8 months now and she has recovered completely. The second child was ventilated for 30 days and had a smooth post-extubation course.

GBS children with respiratory paralysis often are

ventilatory dependent. Despite these facts there are no guidelines based on prospective clinical data to help clinician to decide when to wean from the respirator, these ventilatory dependent patients.[1-3] Therefore, we attempted simple clinical parameters to allow reliable prediction for the appropriate time for weaning these children who were on prolonged ventilation. Weaning duration in patients with GBS is only a function of respiratory muscle force and weaning should never be postponed when improvement of respiratory muscle strength and quality of cough are adequate, despite poor limb force. T-piece was used to train the child to breathe and help in prolonging the duration of spontaneous breathing. This helped us in weaning and deciding the time of extubation. We do agree that the successful extubation was not only due to the use of the T-piece but also due to the natural recovery which occurred during these periods.

Though in 1937 Philip Ayre introduced the T-piece technique for infants and children, Sir Ivan Magill had introduced the concept during World War I.[2] The patient's respiratory system is open to the atmosphere both in inspiration and expiration. Fresh, humidified and warmed gas is connected to the endotracheal tube by one end. The other end is connected to a length of corrugated tubing that acts as a reservoir, allowing the young child to breathe spontaneously with hardly any resistance (Figure 1). The corrugated reservoir connected to the expiratory limb (Figure 1) is sufficiently flushed with oxygen/ air to dilute and wash out carbondioxide. For this, the volume of the expiratory limb should be approximately the same as the patient's tidal volume. An inflow of two and a half to three times the respiratory minute volume prevents carbon dioxide retention. The advantage of using the T-piece properly is that it is essentially free of resistance and thus helps in spontaneous breathing and slow, steady and successful weaning from the ventilator, as in our cases.

In conclusion, long term intubation rather than tracheostomy is possible and probably less traumatic in infants and children, provided portex endotracheal tubes of proper size are used with adequate technical skill and good, dedicated nursing support. Use of T-piece with hardly any resistance in the system helps in the slow weaning process, ultimately resulting in successful extubation, as illustrated by our two cases. However prospective long term studies involving larger number of patients are required.

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