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Clinical Case of the Month

Management of respiratory problems unique to high tetraplegia

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Introduction

In this case a problem common to high level tetraplegics is presented, the insufficient diaphragmatic function due to a spinal cord injury at the C3/4 level. The following case was presented to several colleagues known to be experienced in the treatment of respiratory failure due to high level tetraplegia.

Case presentation

A 63 year old architect sustained, during a riding accident, a fracture luxation of his third and fourth cervical vertebra with intraspinal bleeding, resulting in a complete tetraplegia below C4, incomplete below C3. There were no other injuries. The rescue team found the patient still breathing spontaneously, but severely dyspnoeic. The situation necessitated immediate intubation and controlled ventilation at the site of the accident. The patient was admitted to hospital, a ventral spondylodesis between C3 and C4 was performed, and due to insufficient spontaneous breathing, a tracheotomy followed, through which the patient has since been ventilated. Sonographic and radiological controls of the diaphragm showed active mobility of the left side with no mobility on the right. There has been no further neurologic recovery. The patient is – 4 months after his injury – breathing spontaneously for 4 h a day, otherwise dependent on artificial respiration.

Please discuss your management of this patient.

Management I

GA Baer, MD

A tracheostoma is a socially disabling insult from a layman's point of view, especially when associated with ventilating tubes and the need of intermittent tracheal suction. A phrenic nerve stimulator (PNS) sets the patient free from ventilating tubes, most certainly from tracheal suctioning, and improves the ability to speak. Because of its small size and simple, non-injuring connection to the patient, a PNS is almost no obstacle to patient transfer by laymen and thus, to social integration. Therefore, I would try to provide the patient with a PNS. A normal phrenic nerve is a prerequisite to PNS. Nerve function is tested by measuring conduction time (latency), which is normal below 8 ms and should not be above 13 ms for PNS in adults;[1] values vary with age in children.[2] Normal conduction times do not imply sufficient muscle mass for diaphragmatic contraction. Muscle mass is tested by measuring the shift of each hemidiaphragm on supramaximum tetanic stimulation of the nerve in the neck, which should be 4 cm in adults.[3] The height of the M-wave in the EMG is an unreliable measure. Measuring only the diaphragmatic shift might give a normal result with a deteriorating nerve, which might lead to a disappointing result at later implantation. The patient under discussion obviously has insufficient diaphragmatic function on the left with no function on the right. If his left side function were normal, he should be able to breathe for longer periods than 1 h or more frequently than four times during daytime. Spinal cord injuries around C3 and C4 are frequently associated with damaged motor neurones of the phrenic nerves. Normally, fibres down to C6 join the nerve[4] which is one reason to implant the electrodes preferably intrathoracically.[3] If conduction times are normal, the measurements should be repeated at monthly intervals until it becomes clear whether nerve function improves or deteriorates. The results of the shift measurement on the left side should be compared to that at the beginning of a period of spontaneous breathing. If all results are within normal limits and the shift at supramaximum stimulation on the left is

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remarkably larger than the contraction during spontaneous breathing, implantation would be worthwhile on both sides. Long term full-term diaphragmatic pacing[5] or phrenic nerve stimulation[6] is possible after conditioning of the diaphragm muscle.

Interference with spontaneous breathing on the left could be avoided by slight hyperventilation via PNS, or, even better, by showing the patient the green light of the controller that shines during inspiration. After several days the patient synchronises without any conscious effort.

A danger of asynchrony, however, exists during sleep between the PNS and the patient's centre for opening of the upper airway at inspiration. This can lead to airway obstruction[7] with possibly fatal outcome.[8] The problem might be solved by employing a pulse oximeter with an alarm noise set as high as possible. Perhaps a more reliable solution would be to keep the tracheostoma open at night times, and closed during waking hours. This is most conveniently done by replacing the tracheostoma tube with a Teflon stent with a day time plug.[3] The latter does not inferfere with chin-controlled switches.

Multi-centre studies on diaphragm pacing[9] and on PNS[10] for respirator-dependent patients (RDP) report on 477[9] and 64[10] cases, but there are only two controlled studies comparing PNS with mechanical ventilation for RDP in spinal cord injured (SCI) patients.[11,12] In both studies, there is a trend in favour of PNS. However, the number of cases from each of the two centres is too low for significance. It would be helpful for all engaged in the field if the results of treatment of SCI-RDP were published in a standardised form. Recordings should include diagnosis and demographic data, profession, and social integration before and after the insult and at the end of general rehabilitation and frequency of respiratory infections per year. If PNS is used, the neurophysiological data before implantation, the delay from insult to implantation, the mode and duration of conditioning, and the final result in stimulation hours per 24 h should be given; and if stimulation is less than 24 h/24 h, the reasons should be explained. Such prospective recording of standardised data would provide sufficient numbers of cases for multi-centre studies with significant results on different ventilation modes for SCI-RDP.

Management II

G Exner, MD

Even 4 months after accident recovery is possible. So a final decision to further treatment should be given after 6 months at the earliest. Up to this time we would continue the weaning to prolong the time to spontaneous breathing. Additionally intensive chest-physiotherapy is necessary to improve chest-mobility. In this case of a well-trained 63 year old man it should be possible to get this patient independent from the respirator over the day. At night artificial ventilation should be performed. Over the day the tracheostoma should be closed with a stent, so the patient is able to speak normally. At night a monitoring system is necessary, we prefer a pulse-oxymeter. Nowadays it is quite normal training mobility in a wheelchair (electrically powered) with a respirator useful in emergency cases so the patient has greater independence and, maybe, the possibility to leave the hospital well equipped. On the other hand the implantation of a phrenic-nerve-stimulator could be discussed if the nerve is damaged. In this special case and if recovery leads to a normal function on one side an implantation of the other side is possible theoretically. We have only a little experience about that method. So a bilateral implantation should be performed all the more as supporting the weak function of the working nerve will be good. To compare both methods there are no imperative medical indications giving preference to one of them. Both are good treatments concerning long-time respiration in high tetraplegia. Nevertheless there are differences concerning quality of life and in the ADL. Nursing affairs could be done more comfortably. Requirement of time and personnel could be reduced. Transfers and mobilisation are safer because of small and well fixed devices. Lastly we think from our own experience with 15 patients that the latter method could be proposed to the above mentioned individual.

Management III

U Bötel, MD

A fracture dislocation between the third and fourth cervical vertebral body always means a serious vital menace for a 63 year old patient, as in cases of coincident degenerative alterations the vulnerability of the spinal cord is increased. Hereby only rarely a total crushing of the cervical cord can be found, but as in this case mainly central bleeding in the cord may be noted, whereas the long tracts may be preserved and some regeneration can be observed later on. According to the case in question a complete tetraplegia below C4 is present, incomplete below C3, also pointed out by the initial development, as immediately after the accident spontaneous breathing was present. According to the adopted classification the spinal cord injury must be classified in group A with ZPP (zone of partial preservation), meaning, that also in the further future a total restoration cannot be expected and the classification will remain in group A, but changes in the zone of partial preservation may be observed, more so, as the loss of spontaneous breathing occurred secondarily.

For good reasons an immediate stabilisation by ventral spondylodesis between the third and fourth cervical body had been performed, but the notes do not inform, whether coincidentally an ultrahigh dosage methylprednisolone therapy according to NASCIS II had been administrated. Also a further ventilation by early tracheotomy had been installed apparently.

Although the conditions for elderly patients with high tetraplegia are not very promising, apparently active movement of the diaphragm of the right side reoccurred, enabling spontaneous breathing for 1 h. In connection with a one-sided lung ventilation fatigue will be noted after that time, so that the phases of spontaneous breathing may be increased very carefully. A final prognosis with regard to the total restoration of spontaneous ventilation will certainly not be possible as early as 4 months after injury, so the present treatment should be continued unconditionally. I would not advise the implantation of a diaphragmatic pacer at the present time, but an implantation could be beneficial at a later time, if also no active innervation of the left diaphragm can be observed after the next 3–6 months having in mind, that in case of a one-sided pacer induced ventilation the spontaneous breathing of the other side has to be adapted causing extreme difficulties in many patients. Therefore the therapeutical concept mainly should follow the previous treatment especially as further improvement of the training effects with a sufficient one-side ventilation can be expected.

The prognosis if the tetraplegia is below C4 will be much more unfavourable although with regard to the ZPP some regeneration in the adjacent segments cannot be excluded. Despite the further intermittent ventilation I think, it will be favourable to treat the patient not in the intensive care unit but in a specially equipped peripheral ward, as we have made the best experiences in our department with this proceeding.

Management IV

CP Naumann, MD, ME Baumberger, MD and G Zäch, MD

A lesion at C3/4 with consequent complete sensorymotor tetraplegia below the fourth cervical cord segment represents a special clinical situation, since ventilator support is not always necessary.

In this case, no further neurological recovery occurred 4 months after the injury. In our institution, in addition to pulmonary, otorhinolaryngological and radiological evaluations, a further electrophysiological examination of the phrenic nerve would be carried out to quantify the exact extent of the damage. Based on these results, the subsequent treatment would be planned in collaboration with the patient.

In the meantime, an attempt would be made to improve the patient's spontaneous breathing by specific training of the accessory muscles of respiration and, if possible, instruction in glossopharyngeal breathing.

The following therapeutic options would be discussed with the patient:

(a) Further training of the respiratory muscles to prolong the periods off the ventilator, with the goal of achieving full spontaneous respiration during the daytime. Should this not be possible, or if a tendency to retention of secretions and atelectasis on the paralysed side should become apparent.

(b) Controlled ventilation.
(c) Diaphragm pacing.
(d) Intercostal nerve stimulation.

We would suggest diaphragm pacing (direct stimulation of the phrenic nerve if possible, or by grafting an intercostal nerve to the phrenic nerve).

Diaphragm pacing has significant advantages when compared with other options: fewer chronic infections, better speech and olfaction, greater autonomy, decreased danger from technical ventilator problems, and hence, improved quality of life.

Actual management

B Fromm, MD, G Hundt, MD and HJ Gerner, MD

The patient was admitted to our centre 3 weeks after his injury. Immediately at the site of the accident methylprednisolone was administered according to the NASCIS II scheme. The nasal intubation under fiberoptical conditions was necessary, when a ventral spondylodesis between C3 and C4 was performed. A tracheotomy was carried out after prompt weaning efforts failed. The patient was allowed to breath spontaneously under PSV-mode (pressure support ventilation) with a setting CPAP/ASB (continuous positive airway pressure/assisted spontaneous breathing) of 5/15 mbar and a F_iO_2 of 0,4 (Servo 300 Ventilator, Siemens-Elema, Sweden). Several weaning efforts were carried out, the best result reached 10 min of free spontaneous breathing. The patient developed a bilateral pneumonia. At this point he was transferred to our hospital.

At the time of admission to our centre he was circulatory stable. The patient still suffered from a bilateral pneumonia. He was still breathing spontaneously in the PSV mode. The blood-gas analysis showed no noticeable problems except a hypokapnea (P_aCO_2 30 mmHg). Enteral feeding was managed via a PEG (percutaneous entero-gastrostomy), and the bladder was drained with a suprapubic catheter.

Sonographic and radiological controls showed an active mobility of the left side of the diaphragm and no mobility of the right. The initially measured pulmonary function showed the following values during spontaneous breathing (there was a significant decrease of heart rate and blood pressure but the circulation remained stable) (Table 1).

Although the results of the pulmonary function tests were very low and the blood gas analysis changed rapidly during spontaneous breathing, we started weaning the patient off mechanical ventilation. Weaning trials were carried out in form of progressive ventilator-free breathing (T-piece-weaning), first as

Table 1

Parameter	*Mechanical ventilation*	*10 min spontaneous breathing*
V_T (l)		0.20
RR (b/m)		16
$P_{0.1}$ (cmH_2O)		0.3
RR/V_T (b/l/min)		50
F_{ET} CO_2	29	43
pH	7.45	7.36
pCO_2 (mmHg)	29.4	39.2
pO_2 (mmHg)	137.6	78.2
HCO_3^--(mmol/l)	19.9	21.4
BE (mmol/l)	−1.9	−2.0
SaO_2 (%)	99	94

Table 2

Parameter	*Mechanical ventilation*	*10 min spontaneous breathing*
V_T (l)		0.29
RR (b/m)		13
$P_{0.1}$ (cmH_2O)		2.2
RR/V_T (b/l/min)		35
$F_{ET}CO_2$	25	39
pH	7.47	7.36
pCO_2 (mmHg)	24.3	36.4
pO_2 (mmHg)	94.3	100.2
HCO_3^--(mmol/l)	17.3	20.3
BE (mmol/l)	3.7	−4.0
SaO_2 (%)	99	96

'once daily trials of spontaneous breathing', beginning with 15 min daily. With this mode the patient succeeded in breathing spontaneously for 2 h a day at his best. Beyond this he was not able to make any durable progress over several weeks. We therefore decided to change the timing of his breathing pattern to 'intermittent trials of spontaneous breathing', again beginning with 15 min three times daily. After several weeks of continuous increase of the respirator free breathing intervals, the patient succeeded in breathing spontaneously three times a day for 2 h. After that his pulmonary function showed the following values (with stable heart rate and blood pressure values) (Table 2).

Beyond this he made no further progress over several weeks. We had to re-examine our concepts, considering the implantation of a phrenic nerve stimulator as well as leaving the patient respirator-dependent with intermittent spontaneous breathing periods at daytime and controlled mechanical ventilation overnight. We discussed the problems with the patient. As he was in a very depressed state at that time, he refused any further respiratory training programme, even any further therapeutical steps. That was the reason to transfer the patient to a regular ward, where we expected him to go forward in weaning from mechanical ventilation. Now, 10 months after the accident, he is breathing spontaneously on average 8 h during daytime, and is on continuous mechanical ventilation at night times.

Discussion

Patients with high level tetraplegia generally suffer from respiratory failure. Depending on the level of paralysis and therefore on the impairment of the neural control of the diaphragm, three different groups can be identified:

(1) Patients with no function of diaphragm and accessory muscles (level of paralysis higher than C3).
(2) Patients with partial function of the diaphragm and accessory muscles (level of paralysis C3/C4).
(3) Patients with complete function of the diaphragm and of the accessory muscles in parts (level of paralysis lower than C4).

Patients of the first group will be dependent on mechanical ventilation for the rest of their lives if an implantation of a phrenic nerve stimulator (PNS) is not carried out. Patients from the third group can be easily weaned off mechanical ventilation. The main problems are therefore patients with partial function of the diaphragm and accessory muscles. In general, patients with SCI below the C3 level can be weaned off mechanical ventilation, but this implies a complex and long-term weaning process which may last days, months or even years.[13,14] Considering the patients' self-determination and independence, the implantations of a PNS may be an alternative to continuous or intermittent mechanical ventilation after weaning had definitely failed. As our patient was initially breathing spontaneously and continued to improve, the implantation of a PNS was not our primary aim of treatment up to now. In the following, we would therefore like to point out some of the numerous problems to be encountered in the weaning process of a patient with high level tetraplegia:

(1) Preconditions.
(2) Airway management.
(3) Monitoring.
(4) Time pattern of progressive-ventilator-free-breathing (PVFB, T-piece-weaning).
(5) Quality of life.

Preconditions

Precondition for weaning an individual with high-level tetraplegia off the ventilator is a consenting patient with at least some – even unilateral – diaphragmatic mobility. For successful weaning, this active diaphragmatic mobility should exceed half the maximum diaphragmatic shift of 4 cm during inspiration. This can be examined either by ultrasound or radiologically via fluoroscopy. The lungs and respiratory system have

to be free of serious pathologic findings (pre-existing diseases, injuries or secondary trauma changes), and regular chest physiotherapy with therapists specialised in this field is mandatory.[15] Cardio-circulatory as well as intestinal functions should be stable with physiologic parameters.

Airway management

In many cases of high level tetraplegia, the inability to mobilise tracheal secretions and not the ventilation itself carries the highest risk of secondary pulmonary infections. A tracheostoma undoubtedly is socially disabling. However, as tracheal suction is easily accomplished this way, it is the only way to minimise the risk of secondary pulmonary infections caused by retention of tracheal secretions. Secondly, patients with high level tetraplegia are mostly long-term ventilator dependent and need a tracheostoma for mechanical ventilation and weaning.[16] There are, however, no data available which allow to decide prospectively which patient needs a tracheostoma. It is our opinion that a tracheotomy is only indicated in cases of patients who are unable to be weaned from mechanical ventilation under nasal intubation. Therefore a short term weaning trial via a nasal tube should be carried out, in some cases even on two occasions. Only if this procedure has failed, will we immediately carry out a tracheotomy.

Mechanical ventilation and weaning via a tracheostoma do not exclude the patient's ability to speak. There are several tracheal cannulas available which allow ventilation and speech (eg Tracheoflex Phonetic, Willi Rüsch AG, Kernen, Germany, or Shiley, Mallinckrodt Medical Inc., Irvine/CA, USA). We are therefore very reluctant to close a tracheostoma even after the patients have been weaned successfully from mechanical ventilation.

Monitoring

During the early stages of weaning, it is imperative to extensively monitor heart rate (HR), invasively measure the blood pressure and central venous pressure, and control the respiratory rate (RR), tidal volume (V_T), end tidal carbon dioxide ($F_{ET}CO_2$) and oxygen saturation (SaO_2) via online measurements to avoid cardiorespiratory distress. These measurements (and the treatment of cardiorespiratory distress) are only possible in an intensive care unit (ICU). Later, when the patient remains stable during longer weaning trials, he may be transferred to an especially equipped regular ward, although our experiences do not definitely confirm this proceeding.

There are few predictive indicators which act as a guide to successful weaning trials in high-level tetraplegia. It is generally accepted, but not proven, that candidates fit for weaning should have a forced vital capacity (FVC) of at least 1000 ml and a negative inspiratory force (NIF) more than 30 cmH_2O.[17,18] We, however, started – and succeeded – in weaning patients with high-level tetraplegia whose readings differed significantly from these values.

A number of indices as predictors for a successful weaning do exist for general surgical or medical populations in the ICU and may be helpful in deciding if a patient is capable of breathing spontaneously:[18]

Respiratory rate (RR)	<35/min
Tidal volume (V_T)	>5 ml/kg
Forced vital capacity (FVC)	>10 ml/kg
Increase of P_aCO_2 after decannulation	<8 mm/kg
Rapid shallow breathing index (RR/V_T)	>105/min/l
Negative inspiratory force (NIF)	$>15-25$ cmH_2O
P_aO_2 ($FiO_2=0.4$)	>60 mmHg

Reference values of these parameters for patients with high level tetraplegia are not established so far, but we use them first of all as bedside criteria to avoid cardiorespiratory distress, and secondly as weaning criteria. Pulse oximetry is the most important parameter of sufficient oxygenation during weaning trials, but during weaning trials with O_2 substitution, oxygen saturation is not a predictive parameter to determine a successful weaning.

Time pattern of progressive-ventilator-free-breathing (PVFB, T-piece-weaning)

In accordance with other authors,[13,18] it is our opinion that T-piece-weaning is the only way to successfully wean patients with respiratory failure due to high level tetraplegia. There are, however, some questions left open, one of them is the time pattern of this technique. In general, there are two possible time patterns for spontaneous breathing through a T-piece during weaning trials:[14]

Once daily trials of spontaneous breathing
In this mode, the patient is breathing spontaneously once a day without any help from the ventilator as long as he feels comfortable (up to a maximum of 2 h). At the moment when he shows any sign of respiratory distress, he will be placed on controlled mechanical ventilation for the rest of the day. Patients who tolerate 2 h trials once daily are able to be extubated.

Intermittent trials of spontaneous breathing
During these weaning trials the patient is breathing spontaneously with a fixed time pattern, starting from even a few seconds two to four times daily and increasing the ventilator independent time intervals every day by a given time period (minutes to hours) until they are able to breathe without a ventilator for the whole day.

Our patient himself chose the once daily trials, although our own and the experience of other

centres[19] suggest that intermittent trials tend to be superior in weaning patients with respiratory failure due to high level tetraplegia. Our patient was able to breathe spontaneously for 2 h on a once daily mode, but it was impossible to disconnect him definitely off mechanical ventilation as he just did not show any further progress. In this situation, and in order to enable him longer spontaneous breathing periods, we decided to change the mode to intermittent trials. He again started at 15 min ventilator free breathing periods three times daily, reaching three times 2 h after some days. Then again, he did not show any further progress suggesting that in this case the duration of spontaneous breathing was limited at this point.

Up to now it remains unanswered whether 'once daily trials' or 'intermittent trials' of spontaneous breathing are more successful in weaning patients with high-level tetraplegia off mechanical ventilation. Both methods are currently evaluated in a prospective study protocol.

Quality of life

Weaning patients with high level tetraplegia off mechanical ventilation is a difficult and time consuming task. During this time, weaning trials determine all other daily activities. In order to co-ordinate all daytime activities, an exact time schedule has to be fixed for every day. This allows all members of the interdisciplinary team involved in the rehabilitation of the patient to co-ordinate their efforts, and for the patient to develop adequate social activities under his dramatically changed conditions of life.

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