Thrift, not just surviving, with motor neurone disease. The outcome of the first pre-emptive ‘triple-ostomy’

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Abstract
The following report details the multidisciplinary treatment of a patient with motor neurone disease. The patient, who requested publication of this case, is a highly intelligent and distinguished robotic scientist. He was diagnosed with amyotrophic lateral sclerosis in 2017 and his personal approach to his condition has been to use modern technology and all treatment options to maximise his quality and duration of life. After his research, the patient decided that his life would be significantly improved by formation of an elective ‘triple-ostomy’, this being an end colostomy and suprapubic catheter (for continence), and a percutaneous gastrostomy (for nutrition). We report the peri-operative multidisciplinary approach taken with this case, the surgical procedures, the potential risks and the outcome. The patient is delighted with the result and aims to raise awareness that this may be a treatment option in highly selected patients.

INTRODUCTION
Motor neurone disease (MND) is an incurable neurodegenerative disorder, which most frequently affects individuals in their 60s and 70s [1]. It is progressive in nature and always fatal [1]. Current treatment options are limited.

At the direct request of a fully informed patient, a combined surgical procedure was performed to improve current and future quality of life. We are not aware of another such procedure being reported previously. In this case, it was successful and the patient wishes to raise awareness of this treatment option.

It is important to balance the potential risks of this treatment against the potential benefit and emphasise the multidisciplinary team approach required.
involve significant risks, particularly due to the anaesthesia required with his deteriorating respiratory function, and therefore requested that these procedures be performed in a single operation and relatively rapidly before his respiratory function deteriorated further.

It became apparent that no patient in the region had ever requested such a procedure. It seemed reasonable, in a fully informed patient who understood the potential risks, to proceed to put a team in place to carry out the treatment. A truly multidisciplinary peri-operative approach was necessary. Peri-operative medicine describes the practise of ‘patient-centred, multidisciplinary and integrated medical care of patients from the moment of contemplation of surgery until full recovery’ [2]. Prior to surgery, the patient met the three different surgical consultants that would be required for the surgery to discuss risks and benefits and perform informed consent. He and his carers pre-operatively spent time with the stoma team and the ward nursing team in order to plan the care.

A formal anaesthetic pre-operative assessment was undertaken, including spirometry to assess lung function followed by a consultant review to discuss risks and specific plans for post-operative care. The potential for worsening of MND symptoms and the requirement for post-operative invasive ventilation were discussed including the possibility of a tracheostomy and an extended hospital stay. A step down to non-invasive ventilation on the intensive care unit (ICU) was planned and a preoperative discussion with the home ventilation team was engaged.

The combined surgery was performed electively on the 10 July 2018. A general anaesthetic with target controlled propofol and remifentanil was undertaken. Intubation was undertaken with high doses of remifentanil. Muscle relaxants were avoided as their use in MND patients has been proposed to increase the risk of hyperkalaemia, rhabdomyolysis and potential for prolonged neuromuscular blockage [3]. Post-operative analgesia was in the form of an oxycodone patient-controlled analgesic pump and a subcutaneous ketamine infusion to minimise the risks of opiate induced respiratory depression.

The three surgical procedures were performed in a standard manner; each being well-established procedures in normal practise. Initially, the suprapubic catheter was placed. Secondly, the colorectal team established a pneumoperitoneum and the sigmoid colon and rectum were then mobilised laparoscopically as per routine practise. The bowel was delivered via a trephine left iliac fossa incision marked pre-operatively by the stoma team and an end colostomy was fashioned. Thirdly, the upper gastrointestinal surgeon then placed the gastroscope tube. This was performed under direct laparoscopic vision and the stomach was fixed to the abdominal wall by intra-corporeal sutures. The order of procedures was planned as the catheter placement was extra-peritoneal and would fix the bladder out of the pelvis, aiding the laparoscopic pelvic dissection and hence performed first. The gastrostomy was last to avoid any theoretical tension that may have been placed on the gastrostomy by a prolonged pneumoperitoneum. The total operative time was 3 hours and 40 minutes.

Post-operatively, the patient was taken to ICU ventilated to ensure optimisation prior to extubation and allow extubation straight onto bilevel non-invasive ventilation via a facemask. This was rapidly weaned to self-ventilation. This stepwise approach in the setting of ICU ensured optimal return of respiratory function before extubation. The patient spent 1 day in ICU before discharge to a general surgical ward with ICU outreach service support. The patient was discharged on day 15 post-op.

The patient is thrilled with the result of his operation, from his perspective, it has significantly improved his current and future quality of life. At 12 week surgical follow up, there were no issues and he was discharged to the community team. He genuinely sees the triple-ostomy as something which has transformed his life and has absolutely no regrets. Even when ‘massive disability survival’ is the primary outcome, there may be patients for whom this is an acceptable outcome [4].

DISCUSSION

To our knowledge, this is the first time these procedures have been performed in a single operation for the pre-emptive management of MND. Currently, the only treatment options recommended by NICE for MND are riluzole and non-invasive ventilation [5]. A Cochrane review concluded that riluzole likely extends the life of a MND patient by 2 or 3 months [6] and non-invasive ventilation may improve quality of life but only extends duration by 48 days [7]. The current focus of medical care surrounds identifying and treating symptoms and having frank discussions surrounding ceiling of treatment [7].

Whilst the procedure was successful it was not without risk. The individual surgical procedures were routine and the risks specific to each were fully discussed with the patient. In this specific case, it was thoroughly discussed and documented pre-operatively and for this specific patient, the potential benefit justified the risk. Timing of surgery was also important as it involved coordination of multiple teams. It was vital to perform the operation before the patient’s respiratory function deteriorated further.

We have not performed a cost analysis. However, we would anticipate that this single procedure should result in financial savings when the cost of long-term carers is considered.

We hope this report will challenge traditional beliefs surrounding the management of MND and hope it will encourage clinicians to consider the ‘triple-ostomy’ as a treatment option for highly selected patients.

ACKNOWLEDGEMENTS

None

CONFLICT OF INTEREST STATEMENT

No conflicts of interest.

FUNDING

None.

ETHICAL APPROVAL

No formal ethical approval was sought prior to writing this case report.

CONSENT

At clinic follow-up, the patient directly requested that the case be written up and published. The patient is a contributing author. The patient has signed a document giving permission to use the case and for it to be submitted.
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