Essay

Caring for the Terminally Ill: Nursing a Patient with Oculopharyngeal Muscular Dystrophy

Claire O'Neill

University of Essex

Abstract

OPMD is one of the Muscular Dsytrophies (MD); inherited conditions, characterized by progressive weakness and degeneration of skeletal muscles (Lovering et al 2005). OPMD is an autosomal dominant form, of adult-onset, presented by progressive eyelid ptosis and dysphagia, followed by involvement of other muscles of the head and neck, and eventually proximal limb weakness (Emery 1998). This essay demonstrates advanced understanding of the impacts of OPMD on the physiological, social and psychological functioning of individuals, their family and society. The essay identifys the needs for nursing intervention and, informed by current best available evidence and in conjunction with members of the multi-disciplinary team, formulates effective methods of care with OPMD patients to promote comfort and functional ability.

Keywords: Muscular Dystrophy, nursing, OPMD.

Essay

Whilst working on a Gastroenterology ward at the local general hospital, I nursed a patient with Oculopharyngeal Muscular Dystrophy (OPMD). To preserve anonymity of the patient, they will be referred to as 'patient X' or 'the patient' throughout the essay. OPMD is one of the Muscular

Dsytrophies (MD); inherited conditions, characterized by progressive weakness and degeneration of skeletal muscles (Lovering et al 2005). OPMD is an autosomal dominant form, of adult-onset, presented by progressive eyelid ptosis and dysphagia, followed by involvement of other muscles of the head and neck, and eventually proximal limb weakness (Emery 1998). Patient X was a 72-year-old gentleman who had been diagnosed with OPMD 20 years previously. The patient was now bed-bound, virtually paralysed, and had severe dysphagia. As a result of these disabilities the patient had an indwelling tracheostomy, and a Radiologically Inserted Gastrostomy (RIG). This essay reflects on the nursing interventions implemented to promote comfort and functional ability with the patient, as well as demonstrating advanced understanding exploring the multifaceted impacts that OPMD has on Patient X, his family and society.

Whilst eliciting a full medical and social history (see Appendix A) from the patient and his family, one of the main themes that emerged was the severe effects that OPMD had not only on patient X but also the family. Although patient X was aware of a clear family history of the inherited disease, the patient had declined genetic testing for the disorder when he was younger, and only two of his three sons had opted for a diagnostic blood test. This reflects research that has found a huge diversity in reactions from children of OPMD sufferers and their decisions on whether to have genetic diagnostic tests (Bradbury et al 2009). One of the patient's sons had read extensively about OPMD, and often offered clear direction on how it may be best to help his father. Whilst nursing patient X, at times, I remember feeling unsure of whether to follow such advice. However, Cooper (2008) reiterates that in the cases of rare diseases often the carers and families become the experts.

Patient X's positive mental attitude towards the disease was another theme that emerged from his medical and social history. Patient X was always keen to have surgery and treatment that would improve his comfort or functional ability, even if it did not improve the long-term prognosis. This attitude is reflected in the Krause-Bachand and Koopman (2008) study where most OPMD patients sought medical intervention to address their droopy eyelids and swallowing difficulty and expressed thoughts of 'living for today', and 'making the most of their situation'. The only surgery patient X was not happy about having was his RIG insertion, perhaps because the realisation that he would no longer be able to eat food directly challenged his positive attitude towards the disease. Watt and Whyte's (2003) qualitative study found a link between low mood and worsening dysphagia, as a result of a progressive absence of 'decent food' and the association of food with pain. Patient X was

 Essex Student Journal, 2010, Vol.2(2)
DOI: https://doi.org/10.5526/esj136

currently being prescribed mood enhancers, whereas on previous reviews, before the RIG insertion, the patient had described his mood as 'placid' but not depressed. Patient X's mood may have thus progressed from placid to depressed in line with his worsening dysphagia, and as the opportunity for interventions to enhance his quality of life declined.

The Physiotherapy Team suggested that the patient would benefit from wearing a palm-protector on his left-hand, at two hourly intervals, due to a joint contracture. The aim of the regime was to prolong functional use of the patient's hand for as long as possible. Lovering et al (2005) suggest that physical therapies, such as palm-protector regimes, offer the most promise in maintaining the best level of health for the majority of MD patients, due to unlikely advances in gene therapy significantly altering their clinical treatment in the near future. Patient X, however, sometimes expressed mild discomfort when wearing the palm-protector, and concern that being disturbed whilst sleeping at two-hourly intervals was affecting him psychologically. Krause-Bachand and Koopman (2008) found people with OPMD are often affected more by the psychological aspects than the physical aspects of the disease. Further, although Lovering et al (2005) provide evidence that such protectors prolong function, I suggest this is not the same as promoting health. Health is a holistic entity, linked with quality of life, encompassing the multiple dimensions of physical, functional and psychosocial well being (Aaronson 1990). As a result of the patient's complaints, the decision could have been made not to put the palm-protector on at all, however as Horgan (1958) reminds us, until science finds a cure for the MD, the nurses job is to help the patient to cope with the disease and keep as active and alert as possible. Therefore, in consultation with the physiotherapists, taking into account the patient's wishes and available evidence, it was decided that the patient should only wear the palm-protector for four hours a day, with the aim of achieving an equilibrium between the patient being physically active and psychologically well, thus promoting the best level of health possible under the circumstances.

As a result of the patient's malfunctioning pharyngeal muscles, an indication of OPMD, the patient experienced progressive dysphagia (Krause-Bachand & Koopman 2008). The dysphagia had become severe, and consequently large volumes of secretions often accumulated in his trachea and thus required frequent suctioning to prevent aspiration of fluid into the lungs. As a result of this, the patient had an indwelling tracheostomy. Positively, the patient's tracheostomy tube contained an inner cannula, enabling removal and cleaning of this inner tube to prevent blockages (Russell 2005).

DOI: https://doi.org/10.5526/esj136

I cleaned both the inner cannula and stoma site using normal saline as per instructions from the Respiratory Nurse Specialist. Although a literature search yielded no research in the United Kingdom on the incidence of the use of normal saline in this procedure, a review of hospitals in Australia and New Zealand found that the use of normal saline was common practice to prevent infection around the stoma site (Keogh et al 2008). This is surprising as Trundle and Brooks (2004) identify that neither hydrogen peroxide nor normal saline are required for the cleaning of the inner tube and that cleaning a tracheostomy tube with tap water is sufficient. Further, there appears a large amount of evidence which suggests there is no clinically significant increase in the risk of wound infection in wounds irrigated with tap water compared to sterile normal saline (Bee et al 2009; Whaley 2004). Therefore, I would suggest that there is little proven advantage in using saline for cleaning the tube or the site compared to tap water. Indeed, careful attention to the stoma site has been suggested to decrease the risk of infection, and it appears that it is the frequency of the tube and dressing changes as well as cleaning that reduce the risk of infection rather than the liquid used (Yaremchuk 2003).

Scase (2004) suggests that careful cleaning underneath the tracheostomy sponge is required as there is a risk of skin damage from a tube neck plate constantly resting on the peri-stomal skin. Patient X had his tracheostomy site changed daily, with careful attention paid to the skin resting under the neck plate. Serra (2000) recommends that the stoma site be cleaned at least twice daily, and that increased cleansing is dependent on the individual patient's needs and the amount of secretions. Patient X was often noted as having 'large, thick secretions', which would indicate that his stoma site might have benefited from more frequent cleaning. Russell (2005) identified that although a purpose-made brush will assist with removing debris from within the inner cannula, these brushes may then contain micro-organisms, and should therefore be disposed of daily. I did not witness the brush being changed whilst on long-day shifts. As the stoma dressing was due daily, it would have been efficient to dispose of the brush at the same time.

Socially, a tracheostomy may present problems both practically (with eating and communication) and visually (they may look unattractive) (Benner 1984; Barnett 2006). Literature exists on how the psychological effects of a tracheostomy (Barnett 2006). However, there appears little evidence regarding society's perceptions of tracheostomies and how this affects society from a psychological aspect. I noticed that staff would often talk over the patient, and even when they did talk to the

DOI: https://doi.org/10.5526/esj136

patient they failed to place their hand over the tracheostomy which would have allowed a clear response. The current author suggests that people were scared of the tracheostomy. This is particularly socially disadvantageous to patients who have MD as they are not able to use their own hand to cover the tube to propel their voice. I always spoke directly to the patient and placed my hand over the tube to allow the patient to speak, thus limiting some of the negative social effects a tracheostomy presents.

As a result of suspected pleural effusion, the doctors requested a thoracentesis, otherwise known as a 'pleural tap', procedure. It is well documented that patients are worried about pain and the anaesthetic associated with medical procedures, as well as the procedure itself (Mitchell 2000). Therefore, as part of my explanation to the patient on what to expect regarding the thoracentesis, I suggested that that the procedure would be pain free, as a result of a local aesthetic. Coll et al (2004) suggest that it is more beneficial to suggest that such a procedure will be comfortable rather than pain free, as pain is a subjective construct and multidimensional. I have, also, witnessed patients visually and verbally expressing pain during this procedure. Further, it appears widely documented in the research on surgery and similar procedures that giving information to patients and explaining that a painful procedure may be painful reduces the amount of pain that the patient reports, consequently also reducing their anxiety levels (Clements & Melby 1998; Mitchell 2000).

I used well validated communication skills, such as paraphrasing and summarising, to try and maximise the patient's understanding of the procedure (Epstein et al 2004). After describing the procedure to the patient I asked if the patient had any questions, a method found to be effective in achieving patient understanding and thus patient satisfaction (Baker et al 2007). Reflecting on how I explained the procedure to the patient, I could have perhaps written down my explanation, as the air humidifier in his room appeared to make it difficult for the patient to hear at times. Mitchell (2000) argues that, the level of information people need and how they retain it varies, so information provision should be appropriately tailored to individual need. Patient X had been in hospital a long time and had undergone many investigative procedures, therefore he may not have needed as comprehensive an explanation. However, due to documented findings of patient's complaints about insufficient information provision regarding forthcoming procedures it might be good practice to always provide clear, detailed explanations (Gilmartin 2004; Rhodes et al 2006).

DOI: https://doi.org/10.5526/esj136

It is clear that OPMD is a complex disease, with complex physical, psychological and social consequences, for the patient and the patient's family. Through reflection on nursing patient X, I have learnt that the incurable aspect of the disease does not make a nurse redundant in improving the patient's health. A nurse's role, with a patient who has a condition that cannot be treated from a physiological perspective, merely shifts to focus on maintaining and improving the patient's social and psychological well-being. Through multi-professional communication, (with physiotherapists and respiratory specialists in this particular case), a nurse can be the patient's advocate to achieve equilibrium between the different aspects of the holistic entity known as 'health'.

Bibliography

Aaronson, N.K. (1990) Quality of life research in cancer clinical trials: a need for common rules and language. Oncology, 4 (5) 59-66.

Baker, H., Crockett, R., Uus, K., Bamford, J., & Marteau, T. (2007). Why don't health professionals check patient understanding? A questionnaire-based study. Psychology, Health & Medicine, 12 (3) 380-385.

Barnett, M. (2006) Adapting to living with a tracheostomy, Journal of Community Nursing, 20 (1) 4-7.

Bee, T.S., Maniya, S., Fang, Z.R., Yoong, G.L.N., Abdullah, M., Choo, J.C.N., Towle, R.M., Hong, W.Y., Gaik, I.T.C. (2009). Wound bed preparation- cleansing techniques and solutions: a systematic review, 36 (1) 16-20.

Benner, P. (1984) From Novice to Expert: excellence and power in clinical nursing practice. London: Addison Wesley

Bradbury, A.R., Patrick-Miller, L., Pawlowski, K., Ibe, C.N., Cummings, S.A., Hlubocky, F., Olopade, O.I., & Daugherty, C.K. (2009) Learning of your parent's BRCA mutation during adolescence or early adulthood: a study of offspring experiences. Psycho-Oncology, 18 200-208.

Clements, H., & Melby, V. (1998). An investigation into the information obtained by patients undergoing gastroscopy investigations. Journal of Clinical Nursing, 7 (4) 333-42.

DOI: https://doi.org/10.5526/esj136

Coll, A.M., Ammen, J.R.M., & Moseley, L.G. (2004) Reported pain after day surgery: a critical literature review. Journal of Advanced Nursing, 46 53-65.

Cooper, P.G. (2008) When the Patient and Family Are Experts,Will the Nurses Listen? Nursing Forum, 43 (4) 169-170.

Emery, A.E. (1998) The muscular dystrophies. British Medical Journal, 317 991-995.

Epstein, R. M., Alper, B. S., & Quill, T. E. (2004). Communicating evidence for participatory decision making. Journal of the American Medical Association, 291 2359-2366.

Gilmartin, J. (2004). Day surgery: patients' perceptions of a nurse pre-admission clinic. Journal of Clinical Nursing, 13 243-250.

Horgan, P.D. (1958) Muscular dystrophy, RN, 21 42-45.

Keogh, S., Chitakis, M., Watson, K. (2008) Caring for children with a tracheostomy: a national survey of Australian and New Zealand nurses. Neonatal, Paediatric and Child Health Nursing, 11 (1) 10-17.

Krause-Bachand, J., & Koopman, W. (2008) Living with oculopharyngeal muscular dystrophy: A phenomenological study. Canadian Journal of Neuroscience Nursing, 30 (1) 35-39.

Lovering, R.M., Porter, N.C., & Bloch, R.J. (2005) The muscular dystrophies: from genes to therapies. Physical Therapy, 85 1372-1388.

Mitchell, M.J. (2000) Psychological preparation for patients undergoing day surgery. International Journal of Ambulatory Surgery, 8 19-29.

Rhodes, L., Miles, G., & Pearson, A. (2006) Patient subjective experience and satisfaction during the perioperative period in day surgery: a systematic review. International Journal of Nursing Practice, 12 178-192.

Russell, C. (2005) High-dependency nursing. Providing the nurse with a guide to tracheostomy care and management. British Journal of Nursing, 14 (8) 428-433.

Scase, C. (2004) Wound care. In Russell, C., Matta, B. (Eds) Tracheostomy: A Multiprofessional Handbook (p173-186) Cambridge: Cambridge University Press.

DOI: https://doi.org/10.5526/esj136

Serra, A. (2000) Tracheostomy Care. Nursing Standard, 14 (42) 45-52.

Trundle C, Brooks R (21104) Infection control issues in the care of a patient with a tracheostomy. In Russell, C., Matta, B. (Eds) Tracheostomy: A Multiprofessional Handbook (p343-360) Cambridge: Cambridge University Press.

Watt, E., & Whyte, F. (2003). The experience of dysphagia and its effect on the quality of life if patients with oesophageal cancer. European Journal of Cancer Care, 12 183-193.

Whaley, S. (2004). Tap water or normal saline for cleansing traumatic wounds? British Journal of Community Nursing, 9 (11) 471-472.

Yaremchuk K (2003). Regular tracheostomy tube changes to prevent formation of granulation tissue. Laryngoscope, 113 1-10.

Appendix A: Medical and Social History of the OPMD Patient

Reason for Hospital Admission

- Chest Infection due to OPMD and Gastroenteritis.

Medical History

- Diagnosed 20 years ago with OPMD, after signs of OPMD started to appear.
- Patient experienced a gradual deterioration with his mobility, due to OPMD, eventually he ended up in a wheelchair and is now bed bound.
- Gradually worsening dysphagia and ptosis.
- No known allergies.

Current Medications

- Diclofenac, Solpadol, and Paracetamol for aches and pains.
- Adolat for Hypertension..
- Fluoxetine for depression.
- Eye and Ear Drops.

DOI: https://doi.org/10.5526/esj136

Previous Surgery/Procedures

- Right phacoemulsification for removal of lens with lens implants as had bilateral cataracts- patient had been keen to have this.
- Blepharoplasty 'eyelid surgery' for ptosis.
- RIG insertion.
- Urinary catheter insertion.
- Tracheostomy.

Family History

- Patients mother, maternal uncle, one of his three brothers, one of his three sons suffered from OPMD.
- Only two of the three sons decided to be genetically tested for OPMD.
- Patient's mother died at 79, and maternal uncle died at 53, both due to complications of OPMD.
- The son with OPMD, aged 39, doesn't visit as the son finds this too distressing.
- One of the son's who had not been diagnosed with the disease had an interest in OPMD and was an expert on the condition.

Social History

- Previous occupation- engineering buyer, an occupation which he described as a real joy.
- The patient had secured voluntary redundancy in 1996 as had been worried about his financial situation.
- Divorced 15 years ago, but still in contact with his ex-wife.
- Lives alone.
- Non smoker.
- No alcohol.
- No illicit drug use.

Psychological History

- Previous reviews had said the patient had coped well with his symptoms, for example, he

DOI: https://doi.org/10.5526/esj136

used to state that his swallowing was “not too bad” and knew exactly what to eat, and he completed his home exercises.

- Previously described his condition as 'placid' but not depressed.
- Currently expresses that he is depressed, is now on antidepressants as a result.
- Cognition intact.
- Patient had been eager to have surgery for ptosis and cataracts but not for the RIG insertion.

* Certain details have been omitted, i.e. patient D.O.B to preserve the anonymity of the patient

This article is CC BY Claire O’Neill Essex Student Journal, 2010, Vol.2(2)
DOI: https://doi.org/10.5526/esj136