Ptosis (continued)

- To manage ptosis, surgical elevation of the eyelids can be very successful – several procedures are possible and should be tailored to the individual.
- The aim is to preserve symmetry and allow restored vision while still allowing eye closure. As with most procedures, these are best performed by a surgeon with an interest in this form of surgery, often an oculoplastic surgeon.

Anaesthetics

- If someone with OPMD is to be administered an anaesthetic the anaesthetist should be aware of the existing diagnosis and possible complications that could arise because of the condition.

Limb weakness

- Slowly progressive proximal limb weakness is noticed by some patients, usually commencing many years after onset of other symptoms. Physiotherapy and OT advice may be required.

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Muscular Dystrophy UK
Fighting muscle-wasting conditions

Symptoms card

Oculopharyngeal muscular dystrophy (OPMD)

Name ______________________________

Date of birth ____________ **NHS number** ____________

If presenting at an emergency department, contact the neurology/neuromuscular team and respiratory team at:

____

as soon as possible on:

____

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Oculopharyngeal muscular dystrophy (OPMD)

OPMD is a genetic condition that causes muscle weakness in a characteristic pattern. The first sign of the condition is either ptosis or dysphagia. Very slowly, over many years, these problems worsen.

It is useful to explain the words that make up OPMD:

- 'oculo' – the eye muscles (specifically the eyelids) are affected, causing eyelid drooping (known as 'ptosis')
- 'pharyngeal' – the throat muscles, in particular those related to swallowing, are affected (difficulty in swallowing is known as 'dysphagia')
- 'muscular dystrophy' – usually means the condition is genetic, and causes progressive muscle wasting and weakness, both of which are true in OPMD.

Swallowing

- Because of the dysphagia, swallowing difficulties with food and fluids including saliva can be common in OPMD.
- If dysphagia is severe, there is a danger of aspiration, which increases the risk of a chest infection (aspiration pneumonia), which requires prompt antibiotic treatment.
- Mild dysphagia can be helped by suitable attention to the consistency of the diet and by strategies taught by a SALT. Food supplements may be advised by a dietician if there is weight loss.
- In more troublesome dysphagia, because the upper oesophageal sphincter can obstruct weak swallowing, stretching or cutting this muscle often helps, at least for a while. This is achieved either by stretching the muscle with a dilator, or cutting it – cricopharyngeal myotomy.

Swallowing (continued)

- If dysphagia over many years fails to respond to such measures, then alternative methods of feeding can be used. The most acceptable, in the long term, is a gastrostomy.
- This is often achieved by a PEG, or a RIG. These relatively minor procedures can sometimes be carried out as a day case.

Ptosis

- Increasing ptosis may lead to the eyelid covering the pupil and so obstructing vision. In an effort to compensate for this, the forehead muscles become overactive, trying to help lift up the eyelids.
- People with OPMD often adopt a rather characteristic posture with the head tilted backwards to see clearly. However, the eye itself is not affected, and even when the condition is advanced, people with OPMD have normal vision if the eyelids are raised.