

THE BEST TREATMENT FOR SEVERE DYSPHAGIA IN NEUROMUSCULAR DISEASE IS CONSERVATIVE TREATMENT, FOLLOWED BY GASTRIC FEEDING

Sergiu C. Blumen

Director of the Oculopharyngeal Muscular Dystrophy and Neurogenic Dysphagia Service, The Hillel Yaffe Medical Center, Hadera, Israel

In many neuromuscular disorders, dysphagia is a significant feature and sometimes the presenting symptom. In some of them, like myasthenia gravis, dysphagia fluctuates and is amenable to effective pharmacological control. In other cases it is rapidly progressive, as happens in many cases of motor neuron disease. There is still a large group with slowly progressing, permanent, swallowing impairment that is the main target of dysphagia alleviating interventions (DAI). In a series of 34 such patients (Lacau St Guily et al. 1994) evaluated for cricopharyngeal myotomy (CPM) half (17 patients) had Oculopharyngeal muscular dystrophy (OPMD) followed by patients with Steinert myotonic dystrophy (6 patients), mitochondrial myopathies (4 patients), Polymyositis (3 patients) and other diagnoses (4 patients).

The main questions we will address are: When do DAIs become necessary? At what stage are mechanical/surgical interventions mandatory? Which patients will benefit from surgical DAIs and for how long? Do surgical DAIs prevent or delay the possibility of gastric feeding? What information should physicians use in recommending oral versus non-oral feeding in oropharyngeal dysphagic patients?

Most patients require swallowing evaluation and assistance when early difficulties with solid food lead to meal prolongation, social embarrassment, choking or aspirations; in some cases they are diagnosed late, after aspiration pneumonias or weight loss has already occurred. At the beginning, changes of food consistency and swallowing strategy may provide temporary relief. However, as exemplified by OPMD's natural history, sooner or later, all patients will require some form of mechanical DAI, provided they live long enough. These include: esophageal dilations, botulinum toxin injections and CPM with or without laryngohyoid suspension. Ideally, these interventions are useful when dysphagia is solely due to cricopharyngeal sphincter dysfunction and pharyngeal propulsion, tongue and esophageal function are relatively preserved; unfortunately this is not the case for most patients. Diffuse myopathies affect pharyngeal peristalsis, face laryngeal and tongue muscles as well. Many authorities regard lack of pharyngeal propulsion, severe dysphonia and emaciation as contraindications to surgical DAIs. According to one of the largest series, in the short term most patients benefit from DAI, but the rate of recurrence is roughly one third after three years (Coiffier et al, 2006). Unfortunately, available research studies are uncontrolled and inclusion criteria are rather loose; there is no stratification for dysphagia severity and patients who have or have not lost weight were equally included. Neither are criteria defining success clearly quantified. Judging by the implicit inclusion and exclusion criteria stated in most articles, it seems that the large majority of patients who underwent these interventions would not have required gastric feeding (by percutaneous endoscopic gastrostomy - PEG) in the short run if not operated on. It seems that mechanical DAIs are most useful in patients with definite but moderate dysphagia and there is a limited honeymoon between the patient and his improved swallowing. Unfortunately, they can not replace PED feeding and patients with severe disease who lost weight and experienced repeated respiratory infections are poor candidates. Most studies published until today have not documented significant weight gain or cessation of aspirations in those who already experienced these complications. The Cochrane collaboration review from 2007 is unable to recommend surgical DAIs for chronic neuromuscular patients with dysphagia.

Fortunately, the rate of agreement between clinicians on the criteria to recommend oral versus non-oral feeding is high. These include in decreasing importance: (a) amount and frequency of aspirations, (b) accuracy of diagnosis, (c) history of pneumonia, (d) alertness, (e) cough ability, (f) respiratory status, (g) patient wishes, (h) secretion management, (i) recovery prognosis and (j) ability to perform postures/maneuvers (Logemann et al, 2008).