THE BEST TREATMENT FOR SEVERE DYSPHAGIA IN NEUROMUSCULAR DISEASE IS CRYCOPHARYNGEAL MYOTOMY (CPM)

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Dysphagia derived from the Greek *dys* meaning bad or disordered, and phago meaning "eat". In many neuromuscular disorders, dysphagia is a significant feature and sometimes the presenting symptom. Dysphagia can be rapidly progressive, as happens in many cases of motor neuron disease and can present with slowly progressing, permanent, swallowing impairment that is the main target of dysphagia alleviating interventions (DAI). Dysphagia is the most severe manifestation of Oculopharyngeal Muscular Dystrophy (OPMD) and has the most significant impact on quality of life; moreover, life threatening, choking episodes and aspiration pneumonia may complicate it. In early stages, adapted eating strategies and diet modifications may provide temporary relief; however, when disease progresses, more active, mechanical interventions to ease swallowing become necessary; these include esophageal dilatation procedures, local botulinum toxin injections and crycopharyngeal myotomies (CPM).

CPM is one of the surgical procedures that can open the gate for the food passing from the hypopharynx to the esophagus. In a series of 34 such patients (Lacau St Guily et al. 1994) evaluated for CPM, half (17 patients) had OPMD followed by patients with Steinert myotonic dystrophy (6 patients), mitochondrial myopathies (4 patients), Polymyositis (3 patients) and other diagnoses (4 patients). These interventions are not devoid of complications: after CPM a few patients experienced pulmonary complications. Although in many cases their effect lasts a few years, the impact on the nutritional state and psychological well-being of many patients may be dramatic; they finally stop losing weight, choking or coughing during meals, and the threat of PEG nutrition with consequent loss of independence and self esteem is delayed. 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.

CPM can help in paients with: 1.Permanent dysphagia, but will not be recommended for extreme cases. 2. In selective cricopharyngeal dysfunction (UES obstacle) with relatively preserved tongue activity, pharyngeal propulsion, esophageal motility. 3. In relatively slow progression (OPMD, IBM, mitochondrial myopathy, “idiopathic” dysphagia of elderly people), 4. In good general condition for general anesthesia, and 5. It needs the cooperation of Neurolosist and Otolaryngologist.

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, botulinium toxin injections and CPM.

CPM is useful when dysphagia is solely due to cricopharyngeal sphincter dysfunction and pharyngeal propulsion, tongue and esophageal function are relatively preserved; 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 is most useful in patients with definite but moderate dysphagia and there is a limited honeymoon between the patient and his improved swallowing. Patients with severe disease who lost weight and experienced repeated respiratory infections are poor candidates for CPM. The proper timing of these interventions, in neuromuscular disorders and OPMD patients is very important and must be based on deep knowledge about the natural history of the feeding difficulties in this chronic muscle disease. Our conclusions favor intervention and DAIs become necessary when patients unwillingly lose more than 10% of their weight, develop significant dysphonia or experience more than two episodes of aspiration pneumonia or choking. These are likely to occur in heterozygotes at the end of the second decade or during the third decade since disease onset. In emaciated, severely dysphonic or PEG fed patients, CPM brought about, at best, modest, short-term (several months) improvement.