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BRIEF COMMUNICATION

A Case Report of Myotonic Disease and Gastric Bypass and a Literature Review

H. Gohier[1] · P. Cintas[2] · E. Montastier[1] · M. Bertrand[1] · G. Tuyeras[3] · M. Chalret du Rieu[3] · A. Estrade[1] · P. Ritz[1,4]

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Abstract

We report the case of a woman with myotonic dystrophy type 1, followed for 8 years after a Roux-en-Y gastric bypass. Weigh loss was substantial (53% of initial body weight) with functional improvement in spite of the natural course of the pathology. Five other cases have been published and have reported a relatively positive benefit/risk ratio. Precautions are to be taken at the time of anesthesia and follow-up in order to detect possible degradation of muscle function.

Keywords Gastric bypass · Myotonic disease

Individuals with myotonic dystrophy type 1 have an elevated prevalence of overweight and obesity, creating an additional handicap. It is very difficult for them to lose weight. Five clinical cases of bariatric surgery by gastric bypass have been published [1–3].

We report the clinical case of a 52-year-old woman diagnosed with myotonic dystrophy type 1 (MD1) who underwent bariatric surgery 8 years ago and compare this patient with other published cases.

She presented an MD1 whose initial symptoms appeared around the age of 16, with a myotonic syndrome. Difficulties in walking appeared around the age of 42. She did not present obstructive sleep apnea, but there was a minor restrictive pattern of pulmonary dysfunction. There were no clinical signs of esophageal motor disorders or gastroparesis [4]. Metabolically, there was no diabetes, dyslipidemia, or hypertension.

She underwent a Roux-en-Y gastric bypass (RYGB) in January 2010. Pre-operative weight was 106 kg (BMI 38.5 kg/m^2). Weight loss was regular, with a 25-kg loss after 9 months, that was not responsible for muscle involvement. The patient presented a myotonia of the upper limbs with no associated distal deficit. In the lower limbs, there was a distal deficit in the antero-external and posterior compartments, the cause of an impossibility of standing on the toes or on the heels. The deficit was given a score of 4/5 on the MRC scale.

Eighteen months after surgery, the patient had lost 40 kg. She presented a slight aggravation of walking difficulties. The deficit in the antero-external compartment was stable and was present in extension and flexion of the fingers. The upper limbs still presented myotonia.

Two and one-half years after surgery, the patient had lost a total of 50 kg. The myopathy remained stable with a similar deficit. The axial musculature remained untouched.

Seven years after surgery, body weight was stable around 50 kg for a BMI of 18.2 kg/m^2. The deficit was still 4/5 in the antero-external compartments and for flexion of the fingers, with associated axial motor involvement because cervical flexion was 3+/5. There was no trouble in swallowing. However, the patient ceased all physical activity because of asthenia.

Eight and one-half years after surgery, body weight was stable (53% loss of initial weight). Muscle testing showed that the deficit of the anterior compartments of the leg was stable, as was plantar flexion of the ankle. Anterior flexion of the neck was 4/5. Flexion and extension of the fingers were scored at 3+/5. There were no problem in swallowing and no suggestion of gastroparesis [4]. There was a slight myotonia. The patient had no complaints concerning respiration. The functional respiratory examination remained stable with FEVS at 2.1 L, FVC at 81% of theory, and FEVS/VC at 82%.

✉ P. Ritz
patrick.ritz@wanadoo.fr

1 Nutrition Department, CIO and CHU, Toulouse, France
2 Neurology Department, CHU, Toulouse, France
3 Surgery Department, CIO and CHU, Toulouse, France
4 Inserm U1027, University Paul Sabatier, Toulouse, France

Concerning cardiology, there was an atrial arrhythmia and left ventricular ejection fraction was 50%. The ONSL score remained stable, and there were fewer falls.

In summary, this is a case report of a 52-year-old woman who had presented with a MD1 and who underwent an RYGB. Eight and one-half years after surgery, the result is positive, with body weight stable around 50 kg (BMI of 18.2 kg/m^2). Surgery resulted in functional improvement of the patient in spite of the natural course of the pathology, with no progression of the ONSL score which can be considered as a benefit, with no long-term complication imputable to weight loss.

The literature review comprised five cases with follow-ups in the range of 2 to 7 years [1–3]. All the surgeries in the publications were RYGB, with no anesthesia accident reported. Weight losses were 20 to 41% of initial weight and 53% in our case, with no failure of this strategy reported at any time after surgery. Three-quarters of patients stopped nocturnal ventilation and two cases had no sleep apnea. In the qualitative study of Abel et al. [3], the authors state "Although all patients reported physical and mental improvements, they also experienced that their neuromuscular disorder did not change with the operation. Being more active was possible due to the loss of weight, not because their muscles directly benefited of the operation."

In conclusion, taking usual anesthetic precautions in this pathology [5], and with annual surveillance, surgery for obesity does not seem to aggravate the pathology in these patients and leads to a significant loss of weight with no failure reported, this weight loss results in improved mobility. Special care is recommended as cardiac conduction, gastroparesis, endocrine/metabolic changes, CNS, and respiratory function may be altered with the disease [5]. The maintenance of physical activity is of paramount importance, guided by a specialized team as careless muscle strengthening may increase myotonic symptoms.

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