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Case study

Laparoscopic sleeve gastrectomy in an adolescent with Prader-Willi syndrome: psychosocial implications

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ABSTRACT

Prader-Willi syndrome (PWS) is a complex genetic disorder and represents the most common genetic cause of life-threatening obesity in childhood and adolescence. The indication for bariatric surgery in children and adolescents with syndromic obesity is still controversial. This case report deals with the preoperative medical and psychosocial evaluation of a 16-y-old male adolescent with PWS who underwent sleeve gastrectomy. Information on a 6-mo follow-up is also reported. The preoperative body weight was 223 kg (body mass index [BMI] 80.9 kg/m^2). Comorbidities included severe obstructive sleep apnea with nocturnal respiratory failure, hypertension, and impaired glucose tolerance. At 2- and 6-mo follow-ups, the percent excess weight loss was 16 (BMI 71.8 kg/m^2) and 29.2 (BMI 64.6 kg/m^2), respectively. Comorbities did improve. Intellectual disability of genetic origin per se may not represent an absolute contraindication to bariatric surgery if adequate and tailored clinical and psychosocial support is provided.

Introduction

Prader-Willi syndrome (PWS) is a clinically and genetically heterogeneous disorder owing to anomalies in the 15q11–q13 region. Common clinical manifestations include diminished fetal activity, hypotonia, cognitive impairment, short stature, hypogonadotrophic hypogonadism, and early-onset compulsive hyperphagia leading to severe obesity [1,2]. PWS represents the most common genetic cause of life-threatening obesity in childhood and adolescence. Insatiable appetite, food seeking, lack of satiety, and concomitant intellectual and behavioral difficulties make dietary restriction and standard weight loss programs poorly effective [3]. As a result, most patients experience early-onset serious comorbidities, including obstructive sleep apnea (OSA), hypertension, diabetes mellitus, and heart failure, potentially leading to premature death if not effectively treated [3,4].

Bariatric surgery currently is considered the most effective strategy to achieve sustained weight loss in adults with severe obesity. More recently, clinical and research data support its use in childhood and adolescence [5,6]. Sleeve gastrectomy (SG) in particular was suggested as a reasonable option in terms of safety, efficacy, and tolerability in this age group, with good results in terms of sustained weight loss, improvement or resolution of comorbidities, and satisfying compliance to follow-up [5,6].

Currently, the question of whether bariatric surgery might represent a treatment strategy for children and adolescents affected by syndromic forms of obesity is still a matter of debate. Major concerns exist regarding the safety profile of bariatric procedures in these patients, the amount of lost weight and its maintenance, the management of comorbidities, the effect on somatic growth and skeletal maturity, and the adequate compliance to subsequent nutritional recommendations, changes in lifestyle, and follow-up visits [5,7]. Recently, Alqatani et al. reported that SG is a safe and effective procedure in patients with syndromic obesity [8]. SG also was found effective in terms of weight loss and resolution of

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comorbidities in a sample of 24 obese young patients affected by PWS [8]. However, the topic remains controversial [7] and differences persist among guidelines regarding surgical indications in monogenic and syndromic forms of pediatric obesity [9]. Although psychological assessment is currently recognized as an essential procedure in bariatric surgery, and a part of the multidisciplinary evaluation, little is known about the psychosocial management and the implications of SG in patients with PWS. The aims of this case report are threefold:

1. Describe the psychosocial evaluation and follow-up in an adolescent with PWS who underwent SG.
2. Acknowledge the relevance of a presurgical comprehensive and multidisciplinary evaluation and the importance of the follow-up in this patient group.
3. Emphasize that intellectual disability per se may not represent an absolute contraindication to bariatric surgery if adequate and tailored clinical and psychosocial support is provided.

Case report

A 16-y-old male adolescent with severe obesity presented for a surgical visit to the multidisciplinary bariatric surgery outpatient clinic. The patient and his mother reported a history of early-onset obesity in the context of PWS. The adolescent had already tried multiple weight loss programs, including hospitalization in specialized clinics. All efforts were eventually unsuccessful.

A tailored multidisciplinary assessment evaluation protocol was planned and involved a case manager nurse coordinator and professionals from different specialties (bariatric surgeon, psychiatrist, internist, endocrinologist, nutritionist, and anesthesiologist). All bariatric candidates undergo a surgical visit followed by psychiatric and psychosocial evaluation to exclude major psychiatric contraindications to bariatric surgery, according to the criteria of the *Diagnostic and Statistical Manual of Mental Disorders, 5th edition* [10,11]. Surgical eligibility is further evaluated by a detailed clinical and instrumental assessment. The multidisciplinary team meets monthly, and on request when necessary, to discuss complex clinical situations.

The patient underwent an initial psychiatric interview. He reported a history of previous outpatient neuropsychiatric treatments since childhood. He still regularly attended his referent child neuropsychiatrist and had recently completed a detailed re-evaluation at the child neuropsychiatric inpatient unit. In this setting, the diagnosis of "mild intellectual impairment and subthreshold depressive symptoms in a patient with Prader-Willi Syndrome" was confirmed. In particular, the patient's IQ was 62 with a compatible profile of adaptive skills. The hypothetical indication to bariatric surgery also was discussed owing to persistent severe obesity (body weight 169.20 kg, body mass index [BMI] 62.08 kg/m^2 at the time) despite previous multiple treatments. Subsequent psychiatric and psychological encounters with the patient and his parents were planned to more deeply investigate the patient's awareness and motivation about the surgical program and the presence of a psychologically supportive environment. Concurrently, we provided the usual clinical and instrumental assessment, in connection with the child neuropsychiatric outpatient unit. The patient presented regularly to all scheduled appointments with the psychiatrist during the following 6 mo. He was lucid; oriented to self, time, and place; and cooperative. His speech was coherent although simple. He did not present delusions nor hallucinations. Subjectively, he described his mood as "low," with feelings of sadness, inadequacy, and distress related to his body image and to his daily limitations owing to severe obesity. As in previous years, he still practiced swimming once a week, but he was no longer able to practice any kind of other physical activity and needed support for everyday life activities such as personal cleanliness. He acknowledged difficulties in controlling food intake, despite his efforts to follow the nutritional suggestions and the support from his family in this sense. He also admitted frequently eating snacks and hypercaloric foods outside regular meals. He appeared critical about these behaviors, which he could only partially control. He reported increasing anxiety, irritability, emotional reactivity, and verbal aggressiveness, mainly at school, which he attributed to the uncertainty of undergoing bariatric surgery. These rage episodes were occasional, manageable, and never required the emergency intervention of a psychiatrist nor the introduction of any psychopharmacologic treatment. During the interviews, the patient strongly restated his motivation for bariatric surgery. He reported realistic expectations in terms of improvement in his quality of life and global health and was supported by his parents. The multidisciplinary assessment confirmed the presence of third-degree obesity (weight 223 kg; height 166 cm; BMI 80.9 kg/m^2), severe OSA with nocturnal respiratory failure requiring nocturnal continuous positive airway pressure support, hypertension requiring pharmacologic treatment (ramipril 10 mg/d), and impaired glucose tolerance. From 2013 to 2015, the patient had been treated with metformin, but after 2015 the drug was discontinued for the occurrence of for diarrhea. From 2010 to 2015, he also received growth hormone treatment, which was interrupted for hyperglycemia. Considering the clinical severity of obesity, his multiple comorbidities, and the failure of all previous weight loss plans, bariatric surgery appeared to be a reasonable and feasible strategy. SG was considered the option of choice for this patient. Multidisciplinary collective encounters were held with the patient and his family to discuss and clarify risks and benefits of the surgical procedure and the need for an adequate postsurgery compliance. The patient was admitted to the surgery unit a few days before the procedure to verify his compliance to the hospital context. He underwent SG with no complications. Daily support and monitoring by the nurse multidisciplinary team case manager was planned, in addition to the usual protocol. Psychological encounters were offered regularly to the patient and his parents. The patient was cooperative and did not display behavioral abnormalities nor required psychopharmacologic treatment during his stay.

At the 2-mo follow-up visit, a 25-kg weight loss was observed (weight 198 kg; BMI 71.8 kg/m^2; 16 estimated weight loss percent [EWL%]), with improvement of blood pressure control (105/55 mm Hg; heart rate 60 bpm), which prompted a reduction of ramipril dose to 5 mg/d. Glycemic control also ameliorated (fasting blood glucose [FBG] levels decreased from 103 to 79 mg/dL and glycated hemoglobin [HbA1c] from 54 to 43 mmol/moL). Insulin resistance also decreased, as assessed by a change in the homeostatic model assessment (HOMA) index from 10.2 to 1.6. At the psychiatric visit, he reported improvement in mood, with almost complete resolution of anxiety and irritability. He talked with enthusiasm about resuming the daily activities he had previously abandoned. These also included physical activities (swimming and exercise in a gym). He described good compliance with nutritional recommendations, facilitated by a significant perceived reduction or lack of sense of hunger and food seeking, as confirmed by his mother. He also resumed regular encounters with the child neuropsychiatrist. At the 6-mo follow-up visit, further weight loss was observed with a BMI of 64.59 kg/m^2 (weight 178 kg) and 29.2 EWL%. Glycemic parameters revealed an FBG level of 80 mg/dL, an HbA1c of 39 mmol/moL, and a HOMA index of 2.4. Antihypertensive therapy with ramipril was further reduced to 2.5 mg/d. He reported persistence of subjective well-being and was continuing his physical

activity. He admitted occasional deviations from nutritional recommendations, with consumption of sweet beverages and snacks when meeting his friends outside home. Although limited, these episodes emphasized the need for careful monitoring and support by the multidisciplinary team and the patient's caregivers.

Discussion

This report described the case of a young PWS patient who underwent SG for severe obesity. Submitting adolescents to bariatric surgery still remains a highly controversial topic, mainly in patients with syndromic forms of obesity, such as PWS.

Adequate treatment of these conditions is challenging for physicians as the most beneficial therapeutic strategy remains uncertain. On the other hand, increasing evidence supports the safety and efficacy of recent surgical approaches such as SG [3,6,8]. In young patients affected by syndromic forms of obesity, a multidisciplinary approach is mandatory and must encompass the hospital bariatric team as well as a broader network of experts in medical, psychological, and social areas. In the present case, the psychiatric and psychosocial evaluation was coordinated by the case manager, who played an important role in connecting different professionals throughout the entire evaluation and follow-up trajectory.

The patient was affected by extreme life-threatening obesity in the context of PWS. All previous weight loss programs had failed. The patient, however, showed a strong motivation to undergo bariatric surgery, which was perceived as a last chance to ameliorate his life. In this determination, he was supported by his parents, who were, however, aware of the possible limitations and barriers posed by PWS. Major concerns being discussed within the multidisciplinary team regarded safety of the surgical procedure, possible complications, and uncertainty about future results in terms of compliance and weight loss.

Results from the first 6 mo of follow-up were encouraging. The patient experienced a much decreased appetite leading to a reduction in food-seeking behavior. This was in line with previous findings from Alqahtani et al. who reported improved appetite control and decreased preference for sweet foods in a sample of 24 PWS patients after SG [3]. SG resulted in a significant and long-term improvement in the level of ghrelin, an appetite-inducing peptide hormone mainly derived from the gastric fundus resected in SG [12]. Ghrelin has been found to be more than four times as high in patients with PWS than in obese controls. Of interest, Fong et al. showed that patients with PWS who underwent SG experienced both a significant reduction in ghrelin postoperatively and a successful excess weight loss of $>50\%$ after 2 y of follow-up [13].

Of note, at the 6-mo follow-up visit, the present patient reported a relapsing consumption of junk food outside the planned meals. The pathogenesis of hyperphagia in PWS is complex and only partially understood [4]. In this specific case, it is reasonable to assume that a significant role was played by the desire to appear as a "normal" teenager by adapting his eating behavior to that of his friends and thus achieving their acceptance. These considerations imply that bariatric surgery represents a first step in the context of a severe multifaceted complex situation, involving biological, cognitive, neuropsychiatric, and psychological variables. Adequate tailored psychosocial assessment and management is therefore highly recommended.

Conclusion

Limitations of this review included its methodological design and the relatively short duration of follow-up. Therefore, it is impossible to draw firm conclusions about the efficacy of SG in PWS. However, this case report supports the feasibility of SG in selected adolescents with PWS. Careful, tailored multidisciplinary evaluation and psychosocial support for the patient and the patient's family is crucial, both in the pre- and postsurgical phase.

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The patient and his parents gave informed consent for their data to be anonymously used for scientific scope according to the policy of our institution.

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