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Case Report

Prader-Willi Syndrome and Bariatric Surgery: A Case of Successful Laparoscopic Sleeve Gastrectomy in a Super-Obese Patient

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Abstract

Background: Prader-Willi syndrome is a genomic imprinting error characterized by short stature, multiple endocrine disorders, dysmorphic features, scoliosis, osteoporosis, mental retardation, behavioral and psychiatric disorders, making its management a multidisciplinary challenge. Case presentation: This case report presents a 27-year-old woman with Prader-Willi syndrome (PWS), a body mass index (BMI) of 79.8, obstructive sleep apnea (OSA) and prediabetes treated with laparoscopic sleeve gastrectomy (LSG), the patient exhibited a marked improvement in (BMI) from 79.8 to 62 within 6 months post-surgery (22% reduction) with no significant complications. Conclusion: This case highlights the potential efficacy of LSG as a surgical intervention for obesity in individuals with PWS.

Keywords: PWS; superobese; LSG; syndromic obesity; extreme obesity

1. Introduction

Prader-Willi syndrome (PWS), which is brought on by a variety of genetic pathways, is etiologically caused by either point mutations or imprinting mutations, the later may be caused by microdeletion or point mutations, PWS affects roughly one in 15,000–20,000 persons, with an estimated 400,000 cases worldwide [1,2]. The systemic nature of the syndrome results in some unique features of the effected patients as they may exhibit some features like short stature, multiple endocrine disorders, dysmorphic features, scoliosis, osteoporosis, mental retardation, behavioral and psychiatric disorders, all these complications combined results in three times higher mortality than non-effected individuals [3], the lack of data and the systemic nature of the disease makes it difficult to identify a single cause of the deaths in this vulnerable populations as most causes identified as sudden or unexpected by caregivers [4].

PWS temporal evolution can be divided into phases each phase present at a certain age group like hypotonia and poor feeding in infancy, and weight gain without increase in appetite starting from 2-4 years of age followed by substantial increase appetite which typically resolve by adulthood, the details can be found on Table 1 [3].

Table 1.

Phase.	Description	Age
1a	Hypotonia, poor feeding and weight gain	Birth to 9-15 months
1b	Steady typical rate of weight gain	9-24 months
2a	Weight gain without significantly increasing caloric intake or appetite	2-4,5 years
2b	Frank hyperphagia and heightened interest in food	4,5-8 years
3	increased hyperphagia, food-seeking behaviors, lack satiety	8 years to adult
4	insatiable appetite resolves	Adulthood

2. Case Presentation

A 26-year-old Saudi woman known case of prader-willi syndrome suffers from extreme obesity, weighing 163 kg and measuring 143 cm in height (BMI 79.8), complicated by prediabetes, obstructive sleep apnea on BiPAP, she had mild intellectual disability. No known allergies, she is surgically free, she is compliant on metformin, no other home medications, leads a sedentary lifestyle, and has unhealthy eating habits. When examined, the patient appears extremely obese and noninteractive. Her vital signs are within normal ranges, her abdomen is soft and lax, and hernial orifices are intact. Her routine lab results are listed in Table 2. A biliary US was ordered, and the results showed liver measuring 18.1cm and mild diffused hepatic steatosis, otherwise unremarkable. An esophagogastroduodenoscopy (EGD) was done and revealed a small hiatal hernia, regular Z-line at 36 cm from the incisors with normal duodenum and stomach, a biopsy was taken from the stomach which showed mild chronic gastritis, and negative H. pylori, metaplasia, dysplasia, and malignancy. An echocardiography reveals a normal right ventricle, a left ventricular ejection fraction of 55%, and no major valve disease.

Table 2.

CBC.	Hgb	13.4
	WBC	7.3
	PLT	234
RFT	Cr	37
	Alb	37
Lipid profile	HDL	1.2 (low)
	LDL	2.9 (High)
Vitamins	Vit D	32 (low)
	Vit A	Within normal range
	Vit E	Within normal range
TFT	TSH	6 (high)
	T4	Within normal range
Others	HbA1c	5.9%
	U/E	Within normal range
	LFT	Within normal range
	Iron profile	Within normal range
	Zinc	0.4 (low)

She had a successful laparoscopic sleeve gastrectomy; on day 0 following the procedure, the patient was extubated, conscious, and hemodynamically stable with a GCS of 15/15. On the first post-operative day, the abdomen was soft and lax, the lab results were ordinary, and the patient began taking water sips. Day 2-3 after surgery, the patient was taking fluids and food orally, showing no complaints, and had a soft, lax abdomen with a clean, dry wound. On day 3, the patient was discharged. Following two weeks she lost 16kg and her weight improves at 147kg with (BMI 70), after one month she lost 2 more kg her weight was 145kg (BMI 69.83), 3 months after surgery she lost 8 more Kg and her weight was 137 (BMI 67), And after 6 months her Wight improves at 129 (BMI 62) and lost 34kg in total.

3. Discussion

A multidisciplinary approach involving pediatricians, endocrinologists, dietitians, and surgeons is necessary for the treatment of obese PWS patients to provide the appropriate care for the complex phenotype of PWS patients. There is no definitive management of obesity in PWS, but collaboration among the specialists can lead to a longer lifespan and a higher quality of life [6]. Weight reduction is a key element in the management of PWS patients as it plays a role in controlling the co-morbidities

and enhances the quality of life of the patients, however despite the diverse options available the majority of them yields little efficacy and no co-morbidity resolution.

One of the methods to manage obesity typically advised to patients is physical activity and behavioral interventions. However, this method is not shown to be effective in PWS patients due to compliance barriers. Another method is to prescribe pharmacological treatment like orlistat (pancreatic lipase inhibitor) which have low efficacy and substantial gastrointestinal side effects resulting in poor compliance, and metformin which can improve the sense of satiety and lower anxiety in diabetic patients with no weight reduction. growth hormone therapy can delay the onset of obesity and have positive impact on other aspects of PWS especially if started at a young age. However, it is not effective in reducing the appetite and can not be used in severe obesity or severe obstructive sleep apnea as in our case [6,7]. GLP-1 agonist has promising results as it shown to be well tolerated among PWS patients and effective in reducing the BMI and waist circumference while having positive impact on glycemic control, it also improves the sense of satiety. However, some studies showed no significant difference in weight reduction compared to placebo [8–10].

The extremely high BMI of our case does not favor the pharmacological treatment, as IFSO 2022 guidelines recommend that patients with BMI higher than 35 to undergo bariatric surgery and assure the safety of bariatric surgery in patients with BMI higher than 70 in general population. Although surgery is controversial in PWS patients, it can show encouraging results, so we proceeded with it which leads us to another decision about what type of bariatric surgery to go with as IFSO 2022 guidelines did not specify the type of surgery in BMI higher than 60. Moreover, Since the initial report of bariatric surgery for a patient with Prader-Willi Syndrome (PWS) in 1974, the most commonly used procedures, Sleeve Gastrectomy (SG) and Roux-en-Y Gastric Bypass (RYGB), have replaced previous, highly malabsorptive or restrictive techniques. (14,15), And, the limited data available in the literature focuses mainly on sleeve gastrectomy, gastric bypass and biliopancreatic diversion and confirms the safety and efficacy of the aforementioned bariatric surgeries at least in the short term as long term follow up was not documented in many studies, little complications were noted except for few reported cases of ventral hernia and single case of bowel obstruction all of which were in BPD [11], Due to the complex clinical presentation and the varying degrees of intellectual disability that interfere with patient compliance, the long-term outcomes and effects of BS for individuals with PWS remain controversial. There is still not enough information about the risk/benefit ratio, long-term sustainability, and effectiveness of weight loss. (14), Although, SG have been reported to reduce hyperphagia and ghrelin levels among PWS patients [9]. Additionally, Alqahtani AR assures the safety of LSG in PWS and efficacy in controlling obesity [13], Also, Recent data indicates that LSG reduces the foreign body associated with adjustable gastric banding (AGB) and its potential complications, while presenting lower nutritional risks compared to Roux-en-Y gastric bypass (RYGB). (16)

a proposed recommendation by Wolfe G is to perform LSG then if necessary, add the intestinal component of BPD-DS by doing bypass later on to minimize the risk of weight regain [11].

4. Conclusion

LSG is an effective treatment to control obesity in adult patients with PWS, However, the long-term effectiveness and complications of LSG and the other bariatric surgeries need further studies in this population.

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