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Case Report

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Laparoscopic Sleeve Gastrectomy in a Morbidly Obese Pediatric Patient With Bardet-Biedl Syndrome

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ABSTRACT

Data on the effect of bariatric surgery for syndromic obesity are lacking. This case report presents the preoperative evaluation and perioperative outcomes of a 7-year-old pediatric patient with Bardet-Biedl syndrome (BBS) who underwent sleeve gastrectomy. The male patient was referred to our department for the surgical treatment of his obesity. His preoperative body mass index (BMI) was 55.2 kg/m^2 (weight, 83.5 kg), and he was above the 99th percentile for age and gender. The patient underwent laparoscopic sleeve gastrectomy. The postoperative period was uneventful. Six months after the operation, the patient's weight decreased to 50 kg (BMI, 28.72 kg/m^2). Weight loss was maintained until 3 years after surgery. Dyslipidemia and nonalcoholic fatty liver disease were significantly alleviated. Laparoscopic sleeve gastrectomy may be a safe and effective treatment for morbid BBS-related obesity in pediatric patients. Further data are needed to confirm the long-term efficacy and safety of bariatric surgery in BBS.

Keywords: Bardet-Biedl syndrome (BBS); Bariatric surgery; Pediatric patient

INTRODUCTION

Bardet-Biedl syndrome (BBS) is a rare autosomal recessive disorder due to dysfunction of the primary cilia [1,2]. This ciliopathy is characterized by retinal dystrophy, obesity, post-axial polydactyly, renal dysfunction, learning difficulties, and hypogonadism [3]. The prevalence of BBS varies between populations, ranging from 1:160,000 in Europe and North America [4] to 1:13,700 and 1:170,000 in the Faroe Islands and Kuwait, respectively [5].

Obesity, a noticeable early feature of BBS, usually develops in the first year of life. The incidence of obesity has been reported to be 72–92%. Effective weight management is vital to avoid metabolic syndrome and other obesity-related complications of BBS. Bariatric surgery is recommended for children with class III obesity having a body mass index (BMI) of greater than 140% of the 95th percentile or a BMI greater than 40 kg/m^2, whichever is lower [6]. It is further recommended for patients with class II obesity having a BMI of 120% of the 95th percentile or a BMI greater than 35 kg/m^2 with an obesity-related comorbidity, such as hyperlipidemia, type 2 diabetes, insulin resistance, obstructive sleep apnea, gastroesophageal

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reflux disease, nonalcoholic fatty liver disease (NAFLD), and orthopedic disease. However, the existing literature on bariatric surgery for BBS patients is extremely limited. Here, we describe the 3-year follow-up of a BBS pediatric patient after sleeve gastrectomy.

CASE REPORT

A 7-year-old male patient (Russian) with BBS was referred to our department for bariatric surgery. The patient's medical history at the time of referral included retinitis pigmentosa with night blindness, exotropia, insulin resistance, dyslipidemia, NAFLD, hypogonadism, polydactyly on both feet, speech delay, cognitive impairment, Blount disease, and dental abnormalities. Both the father's and mother's families had no genetic abnormalities. The patient had severe obesity due to compulsive hyperphagia. His preoperative BMI was 55.2 kg/m^2 (height, 1.23 m; weight, 83.5 kg). For the age and gender, his weight was above the 99th percentile and his height was in the 37th percentile. There were no abnormalities on preoperative endocrine examination (TSH, 1.88 uIU/mL; Free T4, 1.02 ng/dL; Cortisol measured in the morning, 19.3 μg/dL). There was no specific surgical history except for the surgical correction of polydactyly 4 years ago. The patient's parents gave their informed consent after a thorough discussion on possible treatment options along with postoperative quality of life, expected weight loss, and possible complications. After an appropriate preoperative assessment, laparoscopic sleeve gastrectomy was performed without complications. The procedure time was approximately 110 minutes. The greater curvature side was removed using four 60 mm linear staplers starting from the antrum region to the fundus under the guidance of intraoperative endoscopy. On the second postoperative day, no leakage was confirmed by upper gastrointestinal series (UGIS) with gastrografin, and the patient was started on a liquid diet. He was discharged on the fifth postoperative day without postoperative complications. At 3 months after surgery, hypokalemia was observed due to repeated vomiting. Only liquid diet was tolerable according to his parents. There was no passage disturbance or obstruction on UGIS and endoscopy. The symptom gradually improved after conservative management and dietary education. A significant decrease in the patient's BMI from 55.2 kg/m^2 (height, 1.23 m; weight, 83.5 kg) to 29.8 kg/m^2 (height, 1.32 m; weight, 52 kg) was noted 3 months after bariatric surgery. At 1 year and 6 months after surgery, the patient's body weight reached a minimum of 44.8 kg. At his most recent follow-up assessment (36 months after surgery), the patient's weight was found to be 49.5 kg. His overall medical condition was improved substantially. **Table 1** shows the changes in laboratory results before and after surgery. His lipidemic profile and liver function remained stable within normal

Table 1. Interval changes in laboratory results

Duration of follow-up	Baseline	6 months	1 year	2 years	3 years
Total cholesterol (mg/dL)	254.7	187.6	190.5	203.2	209.0
HDL cholesterol (mg/dL)	24	39	49	65	
LDL cholesterol (mg/dL)	207	175	124	119	
Triglycerides(mg/dL)	246	124	96	47	
Fasting glucose (mg/dL)	101	72	68	82	76
Fasting insulin (uIU/mL)	43	3.2			
C-peptide (ng/dL)	5.32	1.57			
HbA1c (%)	4.7		4.4	4.5	4.3
Alkaline phosphatase (U/L)	297	134	138	130	133
Aspartate aminotransferase (U/L)	57	23	24	19	23
Alanine aminotransferase (U/L)	66	15	16	12	10
C-reactive protein (mg/dL)	1.0	3.86	<0.3	<0.3	<0.3

HDL = high density lipoprotein, LDL = low-density lipoprotein.

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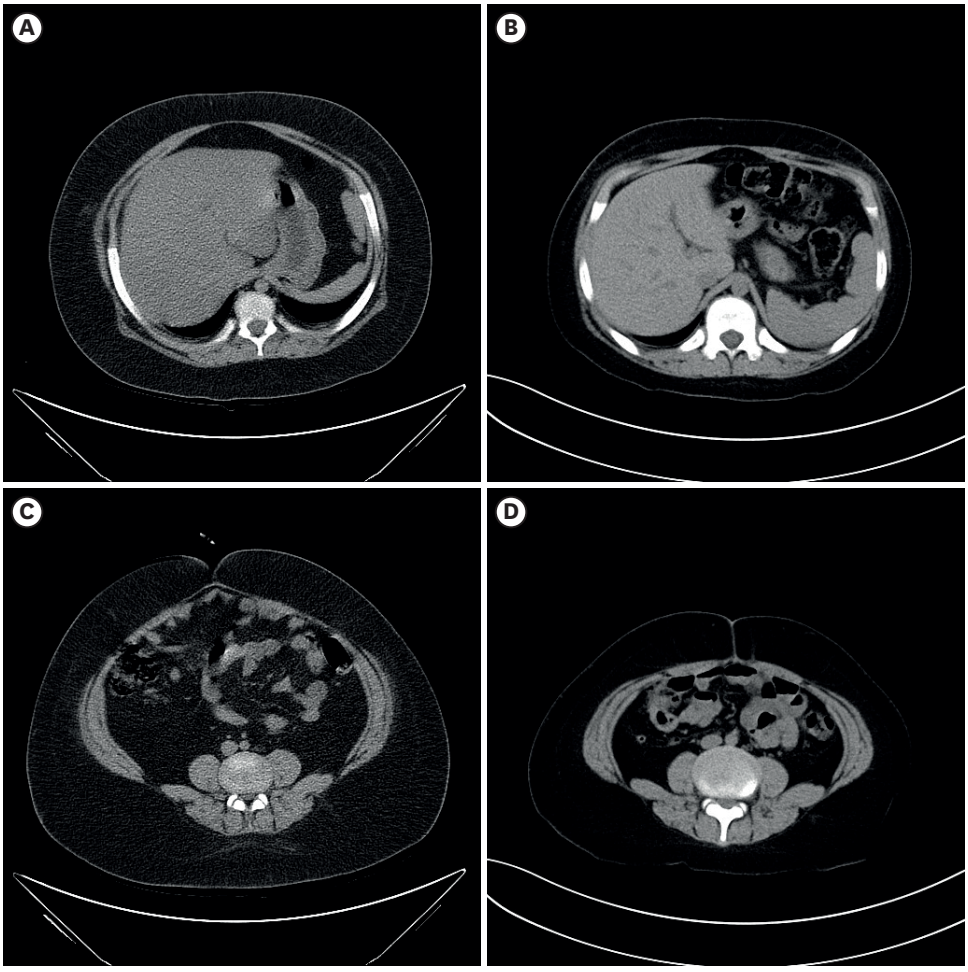

Fig. 1. (A, C) Preoperative CT findings. (B, D) CT findings 6 months after surgery.
CT = computed tomography.

ranges at 6 months after surgery. Fasting insulin (from 43 to 3.2 uIU/mL) and C-peptide (from 5.32 to 1.57 ng/mL) levels were normalized at 6 months after surgery. Abdominal computed tomography showed regressed liver steatosis (from 35 to 65 Hounsfield units) and decreased subcutaneous and mesenteric fat (**Fig. 1**). Immediately after surgery (at 3 months after surgery), height growth seemed to improve to 81st percentile (1.32 m), but it was not good (135.9 cm, 32nd percentile) at the last follow-up (36 months after surgery).

DISCUSSION

The prevalence of obesity in children and adolescents has increased rapidly over the past few decades. Among children and adolescents aged 2 to 19 years in the United States, the prevalence of obesity increased from 1.3% in 1999 to 17.0% in 2014 [7]. In South Korea, the prevalence of obesity among children and adolescents aged 2 to 18 years increased from 8.6% in 2009 to 10.2% in 2016 [8]. Considering the high probability of subsequent obesity in adulthood and the cumulative effect of associated comorbid diseases, the prevalence of severe obesity is expected to increase. Currently, bariatric surgery is considered the most effective strategy to achieve sustained weight loss in adults with severe obesity. More recently, clinical

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and research data support its use in childhood and adolescence [9]. According to the 2019 American Academy of Pediatrics Policy Statement, metabolic and bariatric surgery is regarded as the treatment of choice for children with class III or II obesity with a comorbidity [6].

However, it remains controversial whether bariatric surgery is an appropriate treatment strategy for children and adolescents affected by syndromic forms of obesity. A case-matched study of severely obese patients with and without Prader-Willi syndrome (PWS) by Alqahtani et al. [10] reported that laparoscopic sleeve gastrectomy resulted in similar weight loss and resolution of comorbidities in both groups. On the other hand, Liu et al. found that bariatric surgery could not achieve sustainable weight loss and comorbidity resolution in patients with PWS over a long-term follow-up of 5 to 10 years [11]. There are limited reports on the outcome of bariatric surgery in BBS. A literature review found only two case reports of Roux-en-Y gastric bypass and sleeve gastrectomy for morbidly obese patients with BBS [12,13]. Similar to our report, successful weight loss and alleviation of obesity-related comorbidities were observed after bariatric surgery in a relatively short follow-up period. More studies with long-term follow-up are needed to draw conclusions.

In our case, growth retardation was observed after surgery. Existing evidence suggests that bariatric surgery does not lead to growth impairment, and among older adolescents, several studies have demonstrated that linear growth continues after surgery [14,15]. Therefore, it is necessary to confirm with long-term follow-up whether this is an outcome after surgery or a phenomenon caused by the disease itself.

In conclusion, laparoscopic sleeve gastrectomy is a feasible and promising surgical method for children with syndromic obesity; however, long-term follow-up data are required, especially data related to growth problems. Given the lack of other options for children with syndromic obesity, metabolic and bariatric surgery should be considered, especially when co-morbidities exist.

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