Long term outcome after Biliopancreatic diversion in Prader-Willi syndrome

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Background: Improvement in weight control remains the most important goal of any treatment program in Prader-Willi syndrome (PWS). To date, bariatric surgery experience in Prader-Willi syndrome (PWS) is limited, and different procedures have been used with varying success. Malabsorptive procedures, such as biliopancreatic diversion (BPD), are not always recommended for PWS due to lack of safety data and can involve long-term complications.

Patients & Methods: We report 10 severely obese patients (6 males) with genetically confirmed PWS (7 del15, 3 UPD15) who underwent Scopinaro’s BPD after inability to control food intake with the classical approaches. Surgery was performed on patients aged 18.8±3 yrs. (mean±SD) (range: 15.4-24.4) and the BMI (kg/m2) was ≥40 in all cases (49.9±6.7). At baseline, severe co-morbidities were present, such as obstructive sleep apnea (OSAS), type 2 diabetes mellitus (T2DM), hypertension, metabolic syndrome and/or steatohepatitis.

Results: No perioperative complications were observed. After a follow-up period of 13.9±7.3 yrs. (range 4.8-27; mean age at follow-up: 32.5±6.8 yrs) the maximum weight loss % (MWL%) was 30.7±10 (10.1-52.6). Following BPD, BMI decreased in six patients, stable in three subjects and increased in one individual. The mean BMI at the last visit was 40.5±8.8 (28.9-51.6). After BPD, appetite was reduced in seven cases; eight subjects had hypochromic anemia and seven had diarrhea; OSAS were present in 5 patients and osteoporosis/osteopenia in all individuals. T2DM disappeared and behavioral problems improved in some cases. One patient suddenly died at the age of 37.3 yrs. After surgery all patients received medical therapy to prevent nutritional deficiency.

Conclusion: The long-term outcome of BPD in our PWS seems to be favorable, with a significant reduction of weight excess in the majority of subjects. Thus, BPD seems to be a good option in the presence of severe comorbidity and in selected PWS patients, with cooperating families, when other classical approaches have failed. Due to the presence of specific side effects of the procedure, however, a careful long-term multidisciplinary follow-up is always necessary.

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Biography

Antonino Crino has completed his Postgraduate in Endocrinology, Diabetology and Metabolic Diseases and Pediatrics. He is now responsible for Autoimmune Endocrine Diseases Unit at Bambino Gesù Hospital, Research Institute, Palidoro (Rome). He is Coordinator of the Genetic Obesity Study Group of the Italian Society of Pediatric Endocrinology and Diabetology (SIEDP). He is a referent for central Italy of patients with Prader-Willi syndrome (he follows more than 250 PWS patients). The focus of his research and clinical work is in many fields of pediatric endocrinology. In the last 10 years, he has had a particular interest in genetic obesity, especially in Prader-Willi syndrome. As for the Prader-Willi syndrome, the research programme actually focuses on endocrine problems and on the use and effectiveness of bariatric surgery for treatment of severe obesity in these patients. He has published more than 100 papers in reputed journals and has produced and provided many abstracts both in Italy and at international congresses. He has organized medical congresses and scientific meetings and he collaborates with many scientific institutions either national or international.

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