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Long term outcome after Biliopancreatic diversion in Prader-Willi syndrome

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Abstract

Background: Improvement in weight control remains the foremost important goal of any treatment program in Prader-Willi syndrome (PWS). To date, bariatric surgery experience in Prader-Willi syndrome (PWS) is proscribed, and different procedures are used with varying success. Malabsorptive procedures, like biliopancreatic diversion (BPD), aren't always recommended for PWS because of lack of safety data and might 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 regulate food intake with the classical approaches. Surgery was performed on patients aged 18.8±3 yrs. (mean±SD) (range: 15.4-24.4) and also the BMI (kg/m2) was ≥40 all told cases (49.9±6.7).

At baseline, severe co-morbidities were present, like obstructive apnea (OSAS), type 2 DM (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 followup: 32.5±6.8 yrs) the most weight leader (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 anaemia and 7 had diarrhea; OSAS were present in 5 patients and osteoporosis/osteopenia altogether 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 forestall nutritional deficiency. Conclusion: The long-term outcome of BPD in our PWS seems to be favorable, with a big reduction of weight excess within the majority of subjects.

Thus, BPD seems to be an honest option within the presence of severe comorbility and in selected PWS patients, with cooperating families, when other classical approaches have failed. thanks to the presence of specific side effects of the procedure, however, a careful long-term multidisciplinary follow-up is usually necessary. Recent Publications 1. S Bocchini, D Fintini, G Grugni, A Boiani, A Convertino and A Crinò (2017) Congenital hypothyroidism thanks to ectopic sublingual thyroid in Prader-Willi syndrome: a case report. Ital J Pediatr. 43(1):87. 2. Maltese P E, Iarossi G, Ziccardi L, Colombo L, Buzzonetti L, Crinò A, Tezzele S and Bertelli M (2017) A next generation sequencing custom gene panel as first line diagnostic tool for atypical cases of syndromic obesity: application during a case of Alström syndrome. Eur J Med Genet. 61(2):79-83. 3. Rigamonti A E, Crinò A, Bocchini S, Convertino A, Bidlingmaier M, Haenelt M, Tamini S, Cella S G, Grugni G and Sartorio A (2017) GHRH plus arginine and arginine administration evokes the identical ratio of GH isoforms levels in young patients with Prader-Willi syndrome. Growth Horm IGF Res. pii: S1096-6374(17)30108-9. 4. Brunetti G, Grugni G, Piacente L, Delvecchio M, Ventura A, Giordano P, Grano M, D'Amato G, Laforgia D, Crinò A and Faienza M (2018) Analysis of circulating mediators of bone remodelling in Prader-Willi syndrome. Calcif Tissue Int. doi: 10.1007/s00223-017-0376-y. 5. S Allas, A Caixàs, C Poitou, M Coupaye, D Thuilleaux, F Lorenzini, G Diene, A Crinò, F Illouz, G Grugni, Diane Potvin, S Bocchini, T Delale, T Abribat and M Tauber (2018) AZP-531, an unacylated ghrelin analog, improves food-related behavior in patients with Prader-Willi syndrome: A randomized placebo-controlled trial. PLoS One 13(1):e0190849.

In Prader-Willi Syndrome (PWS), subnormality and compulsive hyperphagia cause early obesity, the co-morbidities of which cause short life-expectancy, with death usually occurring in their 20s. Long-term weight loss is mandatory to elongate the survival; therefore, the dearth of compliance in voluntary food restriction requires a surgical malabsorptive approach. Methods: 15 PWS subjects were submitted to biliopancreatic diversion (BPD) and followed (100%) for a mean period of 8.5 (4-13) years. BPD consists of a distal gastrectomy with a protracted Roux-en-Y reconstruction which, by delaying the meeting between food and biliopancreatic juices, causes an intestinal malabsorption. Indication for BPD was BMI >40 or >35 with metabolic complications. Preoperative mean age was 21±5 years, mean weight 127±26 kg, and mean Body Mass Index (BMI, kg/m2) 53±10. in step with Holm's criteria, all of the topics had a complete score ≥8. IQ assessment was performed in each subject, with a mean score of

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72±10. An arbitrary lifestyle score was given to every subject. Results: No perioperative complications were observed. Percent excess weight loss (%EWL) was 59±15 at 2 years and 56±16 at 3 years, so progressive regain occurred; at 5 years %EWL was 46±22 and at 10 years 40±27. Spearman rank test didn't demonstrate any correlation between weight loss at 5 years and patient data, except with lifestyle score (Spearman r=0.8548, p<.0001).

Current mean age is 31±7 years. Conclusion: BPD needs to be considered for its value in prolonging and qualitatively improving the PWS patient's life.

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