Title
A lower BMI and growth hormone use results in decreased mortality in Prader-Willi syndrome

Authors
Proffit, JN
Osann, K
MacManus, B
et al.

Publication Date
2019

P11.62B
A lower BMI and growth hormone use results in decreased mortality in Prader-Willi syndrome

J. N. Proffit[1,2], K. Osann[1], B. MacManus[3], M. G. Butler[4], V. E. Kimonis[1], J. Heinemann[3], D. Stevenson[5], J. A. Gold[6,7,8]

[1]University California Irvine, Orange, CA, United States, [2]Perinatal Genetics Clinic, Stanford Medical Center, Stanford, CA, United States, [3]Prader-Willi Syndrome Association (USA), Sarasota, FL, United States, [4]University of Kansas Medical Center, Kansas City, KS, United States, [5]Stanford University, Stanford, CA, United States, [6]Dept. of Pediatrics, Division of Genetics and Genomics University California Irvine, Orange, CA, United States, [7]Dept. of Clinical Genetics and Genomics, Addenbrookes Cambridge University NHS Trust, Cambridge, United Kingdom, [8]Loma Linda University Childrens Hospital, Loma Linda, CA, United States

Early mortality has been reported in PWS. Recent studies have shown an increase in survival estimates in the last 20 years. The hypothesis is that it is due to preventative measures to avoid morbid obesity. The PWS Association (PWSA) USA created a long standing bereavement and research program to investigate causes of death and collect data in living individuals with PWS. A familial-response questionnaires from the PWSA (USA), tested the hypothesis that body mass index (BMI), age of diagnosis, clinical symptoms, and growth hormone treatment differ among deceased and living individuals with PWS. Data were available on a total of 2,029 individuals with PWS (114 deceased and 1,915 living) from the USA. Categorical and continuous variables were compared using chi-square and two-group t-tests, respectively. For categorical variables, the effect of age was limited by stratifying for age (<20y v. ≥20y) and birth year (<1994 v. ≥ 1994) and testing with a Mantel-Haenszel test. For continuous variables, the effect of age was limited by adjusting for age as a continuous variable in logistic regression. Average age at death was 31.6 years. Deceased individuals had lower rates of growth hormone use ($p<0.001$) and higher rates of increased weight compared to living individuals. BMI in living and deceased individuals with PWS were 28.6 (SD=11.9) and 51.7 (SD=21.7), respectively ($p<0.001$). This study highlights the benefits of growth hormone, external control of weight, early diagnosis, and the need for a low threshold for bringing affected individuals to medical attention.

J.N. Proffit: None. K. Osann: None. B. MacManus: None. M.G. Butler: None. V.E. Kimonis: None. J. Hei-nemann: None. D. Stevenson: None. J.A. Gold: None.