

Obesity Still Kills

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As I write this, I am working on getting money to pay the funeral of a nine-year-old girl with PWS, and am reminded that PWS is still the deadly appetite. Although many of our children and adults who have Prader-Willi syndrome (PWS) are slim in this day and age due to early diagnosis, education of parents and other care providers, plus growth hormone, I do not want us to get too complacent. Recent calls for help that I have received in our crisis program included: girl 4 yrs, 120 lbs; girl, 5 yrs, 117 lbs; boy 9 yrs, 126 lbs; girl 13 yrs, 254 lbs; boy 11 yrs, 220 lbs; teen 326 lbs; male 24 yrs, 550 lbs;-- and the list goes on.

In studying the 163 PWS deaths that are in our **PWSA (USA) Study of Death** database, Dr. Stevenson wrote: "Regarding the obesity related deaths, in removing all pediatric cases and cases where cause of death was unknown or clearly not obesity related (accidents, choking, etc. – although some of them may have some obesity component), the average weight at death for these individuals was 257 lb." Besides people with PWS being at greater risk of obesity, this obesity puts them at risk to die of obesity related complications. In separate international reports of deaths in PWS, there have been a large proportion (half or more) that were related to respiratory or cardiorespiratory disease. Dr. Linda Gourash (Pittsburgh Partnership) reports in her poster presentation, **Clinical Presentation of Obesity Hypoventilation and Right Heart Failure in Prader-Willi Syndrome**, "The all too familiar

clinical picture of shortness of breath, decreasing physical activity, and leg swelling seen in morbidly obese persons with Prader-Willi syndrome is due to hypoventilation during sleep, leading eventually to respiratory failure with or without right heart failure. The onset may be rapid or slow but is always insidious, that is, initially unrecognized."

Mortality in Prader-Willi Syndrome from New South Wales,(published in Am J Ment Retard. 2006 May) the authors of the study state: "Prader-Willi syndrome is a substantial risk factor for death, above the risk related to intellectual disability alone. Those with Prader-Willi syndrome have higher mortality rates than those with intellectual disability in general. They have a much higher estimated mortality ratio than a comparison group with mild or borderline intellectual disability. The pattern of death in those with Prader-Willi syndrome suggests that obesity-related disease is a likely major risk factor. Management of caloric intake is truly a life-and-death issue for people with this syndrome."

Please remember, and remind your relatives and schools, when you are showing your child love during this holiday season, with Prader-Willi syndrome, you can love your child to death.

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