Human growth hormone therapy in Prader-Willi syndrome

Isabella is a nine-year-old with Prader-Willi syndrome (PWS), secondary to a paternal interstitial deletion (15q-). Her parents seek your advice on the use of human growth hormone (GH) because despite a program of physical activity and rigorous dietary management with the red-yellow-green weight control system, their daughter's obesity is increasing . Lately, she is more tired, has headaches and falls asleep during daytime. Although she was always short, they noticed that her growth is slowing down even further. An Internet search by the parents has suggested that children with PWS may benefit from GH therapy.

LEARNING POINTS

- PWS has been a surveillance target for the Canadian Paediatric Surveillance Program since January 2003, with 42 cases confirmed to date.
- PWS is the most common monogenic etiology of morbid obesity, the consequences of which are the leading cause of life-threatening heart and lung complications and even death.
- The Australian Paediatric Surveillance Unit also confirmed 42 cases of PWS after a two-year period. They established an incidence of genetically-confirmed PWS of 0.4/100,000 children younger than 15 years of age and more than 70% of their cases were diagnosed before 12 months of age.
- Human GH (somatotropin) is used in PWS patients to increase stature and improve the ratio of lean to fat tissue (1,2).
- Global surveillance programs on adverse drug events have identified seven deaths in children with PWS treated with human GH. Common risk factors included severe obesity, a history of respiratory impairment or sleep apnea, or an unidentified respiratory infection (1,3,4).
- A recent review of mortality causes in 27 children with PWS found that none of them were receiving GH treatment (5). Scientific evidence has also identified sleep disturbances that are independent from the obesity in children with PWS (6).
- Although somatropins are not indicated for PWS in Canada (3,7), a physician who decides that their use is in the patient's best interest should consult a paediatric endocrinologist and perform
  - an endocrinological evaluation and anthropometric measurements to prove the need; and
  - sleep apnea studies before and during GH treatment to evaluate the patient's risk of sleep apnea and to detect upper airway obstruction secondary to possible increased growth in lymphoid tissue (1,4).
- The indications and contraindications of this treatment should be discussed with the family.

REFERENCES

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Information on the Red-Yellow-Green Weight Control System is available from The Children's Institute, Nutrition Services, 6301 Nnorthumberland Street, Pittsburg, Pennsylvania 15217, USA. Telephone 412-420-2361, Web site <http://amazingkids.org/main/OrderForm.pdf>. The Canadian Paediatric Surveillance Program (CPSP) is a joint project of the Canadian Paediatric Society and the Public Health Agency of Canada that undertakes the surveillance of rare diseases and conditions in children. For more information visit our Web site at <www.cps.ca/english/cpsp> or <www.cps.ca/francais/pcsp>.