

Helping to prevent obesity and complications in children with Prader-Willi syndrome

Worried parents consult their five-year-old daughter's paediatrician because she is gaining weight rapidly and is not as playful as her classmates. Since school entry, her stubbornness and temper tantrums have increased and she constantly picks her skin. On physical examination, the paediatrician notes generalized obesity with a body mass index of greater than 95% [30 kg (95th percentile), 101 cm

(50th percentile)], a fair complexion, almond-shaped eyes, smaller hands and feet, and hypotonia. Her language is not well articulated. A fluorescent *in situ* hybridization (FISH) test confirms genetically the suspicion of Prader-Willi syndrome (PWS). The challenge for the paediatrician is to coordinate her multifaceted care needs.

LEARNING POINTS

- PWS is due to an abnormality of chromosome 15 with a paternal interstitial deletion in 70% of patients, maternal uniparental disomy in the majority of remaining cases, and less frequently, mutations and other abnormalities of chromosome 15.
- Children with PWS have difficulty in controlling their hunger, which may result in aggressive food-seeking behaviour, food foraging, hoarding and overeating.
- An energy-controlled dietary management program should alleviate the problems of over-eating. Such a program is essential and includes:
 - Early establishment of environmental control and behavioural management;
 - Early introduction of good eating habits with supervision;
 - A concrete dietary management system, allowing for involvement of the child with PWS;
 - Initiation to the red-yellow-green weight control system to structure meal plans and to illustrate food groups and food exchanges for more appropriate self-selection*;
 - Specification of the number of calories in the order of 7 kcal/cm to 8 kcal/cm for weight loss and 10 kcal/cm to 14 kcal/cm for weight maintenance;
 - Reinforcement of exercise therapy with increased and supervised fun activities.
- Medical complications of obesity, such as diabetes, osteoporosis and heart failure, are preventable with ongoing managed care.

*Information on the Red-Yellow-Green Weight Control System is available from The Children's Institute, Nutrition Services, 6301 Northumberland Street, Pittsburgh, Pennsylvania 15217. 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 Health Canada's Centre for Infectious Disease Prevention and Control 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>.