



6th International IPWSO Conference
Cluj-Napoca 21.06.07 - 24.06.07
"Coming here, you will discover yourself, discovering others"
Romanian Prader-Willi Association

ASOCIACIÓN MADRILEÑA
PARA EL SÍNDROME DE
PRADER-WILLI



NUTRITIONAL INTAKE BY YOUNG CHILDREN WITH PRADER WILLI SYNDROME

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INTRODUCTION: Prader-Willi syndrome (PWS) is a genetic disorder associated with hyperphagia that typically occurs from an age between 2-6 years, and without caloric restriction these children will become obese. The families and caregivers of these children receive nutritional counselling, however limited information is available on how families implement the dietary restriction and there is limited knowledge of the actual nutritional intake among young children with PWS. The nutritional intake of 2-, 3- and 4 years old Norwegian children with PWS have been evaluated and compared with the results of the national food surveys for children 2 years and 4 years in Norway.

MATERIAL AND METHODS: We have followed a group of 6 children with PWS. They constitute the total known population of children with PWS in Norway born between 2000 - 2002. All children in this study were diagnosed with PWS in their first months of life. Assessments of the food intake in the age groups 2-3 years and 3-4 years was performed twice a year by structured food interview with parents, carried out by a nutritionist. All interviews were performed by the same person. One assessment was completed using the 24-h recall method via telephone with one of the parents. All interviews, except the 24-h recall, were translated into a typical three day food consumption for each child. For the 24-h recall calculations are made only using information from the food consumption for the 24-hour period. In the age group 4-5 years the parents recorded the children's food consumption for a total of 7 days, divided into two periods during the year, by using a precoded food-diary developed for a national food survey among Norwegian 4 years old children. The recorded data was encoded and analyzed using a food database and software systems developed at the Institute of Nutrition Research, University of Oslo.

RESULTS: We found a mean total caloric intake in the age group 2-3 years: 772 Kcal/day, age 3-4: 862 Kcal/day and age 4-5 years: 953 Kcal/day. This caloric intake amounted to 54 % and 65 % of average caloric intake in the national food surveys for the 2 years and 4 years old Norwegians. When we express the calorie intake for the children with PWS per kg, we found in the age group 2-3 years: 54 kcal/kg - 99 Kcal/kg, mean 72 Kcal/kg, median 70 kcal/kg, in the age group 3-4 years: 48 Kcal/kg - 76, Kcal/kg, mean 65 Kcal/kg, median 68 Kcal/kg. And in the age group 4-5: 49 kcal/kg - 76 Kcal/kg, mean 64 Kcal/kg, median 67 Kcal/kg.

The mean percentage of energy from fat in group age 2-3: 25 E% (range 15-39 E%), age 3-4: 24 E% (range 19-32 E%) and age 4-5: 25 E% (range 20-30 E%).

The mean percentage of energy from sugars in the group age 2-3: 4,3 E% (range 0,6-20,4 E%), age 3-4: 2,6 E % (range 0,5 -6,4 E%) and age 4-5: 4,9 E% (range 0,5-13,4 E%). In the national surveys the mean percentage of energy from sugars were found to be 11,7 E% and 15,1 E% among the 2 years and 4 years old.



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CONCLUSION: Children with PWS consume fewer calories than reference values for age and gender, even though a variation in calorie consumption is seen within the PWS group. Children with PWS consume less fat and less sugars than the average among other Norwegian children their age and with few exceptions their intake of fat and sugars are within the general recommendations of max 30 E% from fat and max 10 E % from sugars. An evaluation of the children's intake of micronutrients is required to make sure their nutritional intake is optimal for growth and development.