

‘The more mobile Melanie became, the more she would go hunting for food. I remember how she stood on tiptoe and pulled the bread off the stove and how she went for the jam in the fridge. At the beginning, she was still slim and we watched with some amusement. At nursery school, she started eating the bread other children had brought.’

Weight and Energy Balance

Parents of children with PWS have to deal with feeding problems shortly after the birth, although initially they are the opposite of later problems. As infants, PWS children are underweight. At this stage, the desire for them to put on weight is the main concern for their parents and the doctors looking after them. It is important to feed infants and babies well and to find the time to do this. When PWS is diagnosed early on, many parents are so wor-

ried about their child's anticipated obsession with food that they do not feed their child enough in the first few years and thereby aggravate their growth problems. The risk is heightened by the fact that PWS infants tend to remain quiet and do not scream when they are hungry. It is therefore helpful for parents to consult a nutritional adviser early on in order to find out how much their child should be eating and drinking each day if they are to achieve optimal nourishment.

All parents whose child has been diagnosed with PWS know that, at some point, their child is going to develop an enormous appetite. The change in eating behavior usually begins between the ages of two and four and does not occur overnight, but gradually. This transformation is extremely confusing for the parents, who are naturally concerned about their child's eating habits. Is the child really over-eating? Is his/her appetite under control? Consulting a nutritional expert can also be very helpful at this stage.

The constant urge to eat causes PWS children to become massively overweight if nothing is done to stop it. They particularly love to eat sweets and other

■ [Fig. 8] The change in eating behaviour usually sets in at an age between two and four years. It never occurs overnight, but always gradually. The girl is crawling into the oven to get to the fresh bread, since she is not yet able to walk properly.



high-calorie foods. They will try to get hold of them whenever possible, sometimes employing very cunning ways of doing so. [fig. 8] Since children with PWS are less active and enthusiastic about movement than other children, they also burn off fewer calories. The weight increase that results from the imbalance between excessive eating and reduced energy consumption serves only to dissuade them from exercising even further – a vicious circle. Body fat mainly tends to develop around the thighs, upper arms, abdomen and buttocks, while the lower arms and legs remain slim. Obesity is all the more obvious since children with PWS grow slowly and not very tall.

The reason for the constant need to eat is a disordered satiety function. This is probably the result of a hormone imbalance, although this has not yet been clearly identified. It is thought that one or more signals which indicate satiety do not reach the hypothalamus in people with PWS, so that they never feel full and must always be on the lookout for food. Despite their increased fat mass, their brain tells them that they are always hungry and need to eat constantly. The main energy control point in the hypothalamus does not detect the energy reserves available (the excess fatty tissue), but only sends out the message 'keep eating'. Healthy people find it hard to imagine such a feeling of hunger, but it should not be underestimated. It can totally govern the thoughts and actions of persons with PWS. [fig. 9]



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[Fig. 9] Drawing by the
7-year-old Hirota.
It shows his mother and
his insatiable appetite.

Outside help from doctors and nutritional advisers can take some of the burden off the parents where eating is concerned. Psychological counselling is also helpful in relation to the monitoring and limitation of the child's food intake. This adjustment of eating habits also has an impact within the family: everyone, including parents, brothers and sisters, must abide by certain agreed rules with regard to eating, such as fixed meal times and carefully measured portions. Siblings must not spoil their brother or sister by giving them extra portions or sweets. The whole family must adapt to the exceptional abilities of PWS children to obtain food. In most cases, even the fridge and kitchen cupboards have to be locked. Parents must also bring their child up without using food as a reward.

Since children with PWS enjoy eating and will put anything in their mouths, they are particularly at risk – as are all small children – from poisonous substances in the house and garden. Medicines, cleaning equipment, paint, etc. must be locked away and a close eye kept on poisonous plants in the garden.

As they grow older, new problems arise as the children come into contact with other people: neighbours, friends at nursery and school, teachers and relatives must be made aware that the parents are not stopping their child from eating through maliciousness, but out of necessity. Informing other people be-



■ [Fig. 10] An adolescent with PWS before the growth hormone era.

‘Jeremy, even now at the age of 10, is really bone idle. He is happiest sitting down. He can spend hours doing puzzles, watching TV or just gawping at what’s going on around him.’

comes very important. As they get older, the children themselves can also understand why they have to watch what they eat. However, the need to eat will triumph more and more often over the desire to please the parents (by not eating). People with PWS therefore need their food intake to be controlled throughout their lives by their carers in order to prevent obesity. [fig. 10]

Reduced Activity Levels

Researchers still do not know exactly why children with PWS are so prone to obesity. It is clear, however, that their sense of satiety is underdeveloped and that they could therefore eat endlessly unless someone stops them. It is nonetheless also evident that PWS children burn the same amount of calories as other chil-