

A disease often misdiagnosed

Prader-Willi Syndrome embodies a cruel irony, one that is not lost on the parents and professionals who deal with the disorder.

If Prader-Willi is diagnosed at an early age, then the victim can be spared many of the debilitating affects of the disorder — including retardation, physical disabilities and death.

Yet, even though the disorder was first identified in 1956, many doctors and medical professionals are unaware that Prader-Willi exists. Thus, victims are usually not diagnosed until they reach their teens — when much of the damage has already been done.

"We have a real problem with misdiagnosis," said Rita Welch, president of the Prader-Willi Greater New York Association. "Doctors tell parents that their kids have cerebral palsy or muscular dystrophy or multiple sclerosis or that the child is just slow in developing."

"Most of the people who have been diagnosed as Prader-Willi in the past year have been 16, 17 or 18 years old. And they are diagnosed after parents hear about our association, and they contact us to see if we have an answer about what's wrong with their child."

The symptoms of Prader-Willi include short stature, lack of muscle tone, underdeveloped sex organs, mental retardation and an "endowment" toward temper tan-

trous. — the real common denomi-

nator is an insatiable appetite. That, combined with the Prader-Willi victim's lack of physical development, can lead to obesity of such proportions that it can become life-threatening.

"If we don't control the weight, the average lifespan for a Prader-Willi person is the mid-teens," said Mrs. Welch, whose daughter has Prader-Willi Syndrome. "The overeating is a lifetime problem, and there is no way to control it."

Persons with Prader-Willi crave food so much that they will go to preposterous lengths just to eat, Mrs. Welch noted.

"We had a lock on our refrigerator and our cupboards, but my daughter learned how to pick the locks," she said. "She would take the hinges off the cabinets. She would go to the neighbors and tell them that I was baking and that I needed eggs or butter or whatever she thought she could get from them."

"These kids will eat cat food, garbage — almost anything is edible to them."

The physical problems related to the overeating include diabetes, hypertension, and heart and circulation problems. Moreover, Mrs. Welch said doctors have found that obesity in Prader-Willi persons can cause or increase the brain damage, which is manifested by retardation and behavior problems.

Obesity and the lack of physical/sexual development can also cause a lack of self-esteem and result in additional

psychological problems, she said.

If the child's weight is controlled at an early age, Mrs. Welch said, the brain damage and psychological problems can be kept at a minimum. But a Prader-Willi person who is still overweight as a young adult usually has an I.Q. of about 70 and a temper which can be sparked by the drop of a hat.

The most important aspect of treatment for a Prader-Willi victim, Mrs. Welch said, is weight control. That means a strict diet and constant monitoring, which can be extremely difficult, especially when the behavior problems are considered. Some parents have reported that their Prader-Willi children become violent when deprived of food.

The best means of controlling the diet of a person with Prader-Willi is putting them in a controlled environment, like a group home or residential facility which has the capacity to keep them away from food, she added. But she knows of only two residential facilities in the entire country for persons with

Prader-Willi, and both only serve children under 21. One, Rhinebeck Country School, is in upstate New York. The other is in Washington State.

Moreover, there is a difficulty in getting public officials to recognize the need for special treatment of Prader-Willi victims, Mrs. Welch said.

"I have asked the (Board of Education's) Committee on the Handicapped repeatedly to be aware of the syndrome so that Prader-Willi children can be identified early and placed immediately at Rhinebeck," Mrs. Welch said.

"They say they are looking at it. Giving it thought and consideration.' Meanwhile every one of these children is having difficulties."

"The situation for adults is worse. The adults with Prader-Willi have never been served and they have worse medical problems."

"These people are experiencing a serious medical plight, and there is no place to put them."

— JULIE MACK
Advance Staff Writer