Q&A for GV May-June 09

The following two questions were answered by Dr. Jim Loker who is on our PWSA (USA) Clinical Advisory Board and the father of 14-year-old Anna who has PWS.

Janalee

Vomiting and PWS

Q Can anyone answer for me the reason why our children who have PWS do not vomit? I'm unsure whether it is due to a lack of gag reflex or low muscle tone, or something else. Anyone care to enlighten my why no vomit and why it is so serious if they do?

A Vomiting is actually a complex reflex involving the stomach and central nervous system. Since the vomiting center is located in the hypothalamus, it appears to be affected in PWS as is other hypothalamic activities. Reflux and rumination (food is regurgitated, rechewed, and reswallowed) do not involve the hypothalamus, so they can and often are seen in PWS. True vomiting may (but not always) be a symptom of a more serious problem than in the general population.

Heart Problems and PWS

Q I heard about a child with PWS who has severe dilated cardiomiopathy. Is this common?

A There are metabolic and mitochondrial syndromes that have both hypotonia and dilated cardiomyopathy, but it is not a common feature in PWS. Although it is not common, it is known to happen. In our PWSA (USA) review of deaths, a 17 month old girl died from dilated cardiomyopathy. The Japanese group had two infants in their 2004 study die from dilated cardiomyopathy. These are most likely due to a viral myocarditis and had nothing to do with the Prader -Willi syndrome. The hypotonia and other features of PWS will complicate care of anyone with dilated cardiomyopathy.

The most common heart failure seen in PWS is obesity related right heart failure due to pulmonary hypertension as a result of hypoventilation, obstruction, obesity, etc. Keeping your child with PWS slim is the best way to prevent heart complications.

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