PWS Growth Hormone Precautions Update – 2/11

We advocate a sleep study before the start of growth hormone (GH) on infants, children and adults with Prader-Willi syndrome, and then a follow up study 6-8 weeks later. If there is worsening of obstructive sleep apnea (OSA) on GH temporarily stopping the GH is recommended until the cause is understood. Frequently the OSA can be corrected by removing the adenoids and tonsils or lowering the dose of GH (in the face of an abnormally high IGF-1). We also recommend taking precautions during bouts of upper respiratory infections.

There are reports and discussion in the medical literature about adrenal hypofunction in PWS. Single measures of cortisol levels will not be helpful and adrenal challenge tests may be warranted. Please consult an endocrinologist for their input and advice before starting growth hormone treatment.

Infants with PWS, may have gastroesophageal reflux disease (GERD) which causes obstructive hypopneas/apneas, so if an evaluation is positive for GERD, an anti-reflux medication may be prudent before starting GH.

Studies have shown that in most individuals with sleep-disordered breathing due to PWS, GH can actually improve (or at least doesn't worsen) the apnea (Haqq et al, 2004; Miller et al, 2006; Festen et al, 2006). Withholding GH from those with sleep apnea may be detrimental on several levels, thus monitoring the child with PWS closely when starting GH to make sure that they do not worsen is the recommended approach.

The FDA has a statement warning that there could be an increased risk of death associated with GH due to a recent study in France indicating that there may be a slightly increased risk of death in certain individuals treated with GH. PWS is not one of the groups mentioned as being at increased risk - they specifically mention idiopathic short stature and isolated GH deficiency.

Jennifer Miller, M.D., M.S. – Endocrinologist --PWSA (USA) Clinical Advisory Board
Merlin G. Butler, M.D., Ph.D. – PWSA (USA) Scientific Advisory Board Chairperson
Daniel J. Driscoll, M.D., Ph.D. -- PWSA (USA) Clinical Advisory Board Chairperson

Prader-Willi Syndrome Association (USA), 8588 Potter Park Drive, Suite 500, Sarasota, Florida 34238
800– 926-4797 * 941-312-0400 * Fax: 941-312-0142 * info@pwsausa.org * www.pwsausa.org