

*Here are some questions that have come to PWSA (USA) and answers from members of our Scientific and Clinical Advisory Boards.*

Q: Is low sodium possibly related to the medications? I had a call from the mother of a 31-year-old male. In Sept. he appeared to be seizing and had trouble breathing. He did not test out as having seizures, but was very low in sodium and magnesium. They put him on magnesium and brought it up, but cannot get his sodium above 130. Right now it is 124. They have restricted his fluid intake and salt his food more. He is on a LOT of medications. His behavior is now wonderful, but is his medical problem due to reaction to one or a combination of meds? He is on:

Loratadine 10 mg a day, Oxcarbazepine 600 mg 3x a day, Chlorpromazine 50 mg 2x day, Chlorpromazine 100 mg 2x a day, Buspirone 10 mg 3x a day, Benzotropine 1 mg 2x a day, Fluvoxamine 100 mg 2x a day, calcium 600 mg 2x a day, magnesium 3x a day, plus a multivitamin.

A: (1) You are absolutely correct! His low sodium may be due to the 1) oxcarbazepine, 2) fluvoxamine, and/or 3) chlorpromazine, or a combination of all of the above. Low sodium can cause seizures. It would be helpful to know which meds were added last, as there may be an additive effect leading to the problem. There is also the possibility of endocrine or renal problems causing the low sodium, but I suspect it is the medications. If oxcarbazepine is the culprit but is proving to be very helpful to the behavioral control, Dr. Gourash and I have had excellent success adding low dose Lithium which causes free water loss through the kidneys; the added benefit of Lithium is mood stabilization. This must be done with someone who has experience using Lithium, as there are other side effects. Oxcarbazepine can be used successfully with positive effects on mood and behavior if side effects are monitored and carefully addressed. This fellow is on a lot of meds, increasing the potential for drugrelated side effects.

*Janice Forster, M.D. Pittsburgh Partnership*

(2) Every single person with PWS that I put on oxcarbazepine (Trileptal) developed lower sodium when the total daily dose exceeded about 900-1200 mg. The sodium may stay in the safe range, but it always goes down some. I am certain that the problem with this young man is primarily the oxcarbazepine, but the other medications could be contributing. The physician in charge may wish to adjust the oxcarbazepine dose downward and recheck the sodium with each dose change. The effect is dose related but not necessarily linear. I had this experience with about 12 patients. Even if the sodium is not dangerously low, you have to keep checking it. It can drift down slowly over weeks or months to a dangerous level. Physicians are not used to how sensitive people with PWS are to the effects of oxcarbazepine on the sodium and need to be very vigilant. I feel much safer if the patient is also being treated with Lithium. We also had 2 patients (one had an atypical deletion; the other had an imprinting center deletion) who developed low platelets. One was dangerously low and recovered as soon as we stopped the oxcarbazepine.

*Linda Gourash, M.D. Pittsburgh Partnership*

Q:How common are seizures with PWS? What type? Recently, my granddaughter was diagnosed as having seizures. The last few times were considered as tonic (also called Grand Mal or a Convulsion) seizures, but she did not jerk as I thought tonic seizures characterized. She only dropped to the floor unconscious with muscles restricted. They did not last very long, but it sure was scary. After some testing, we found out that a characteristic she often has is, in fact, a partial complex seizure. When excited and happy, she will cross her eyes, twist her tongue and move her fingers in a strange way.

When this happens, she can be pulled out of it by just saying her name. We always thought this was her way of showing happiness and were surprised that this was considered a seizure. Is this common with PWS?

A: From our PWSA (USA) medical data base of 1,747 respondents:

Ages 0-5 = 6% seizures

Ages 6-18 = 9% seizures

Ages 18+ = 13% seizures

All ages with UPD = 6% seizures

All ages with deletion = 12% seizures

All ages with imprinting = 3% seizures

All ages with translocation = 26% seizures

PWS like or type of PWS unknown = 13% seizures

Seizures are less common than with Angelman, and those with PWS who do have seizures do not often have the classic epileptic grand mal seizures. Many just have subtle changes in behavior as with your granddaughter or just phase out for a while, so many seizures may go undiagnosed.