

Name: \_\_\_\_\_

Score: \_\_\_\_\_ # Wrong **Pass / Fail**  
(A passing score is 11 wrong (75%) or higher)



## Health Highlights for Caregivers of Persons with Prader-Willi Syndrome

### ANSWER KEY

#### Match the correct definition

- |       |                                 |   |
|-------|---------------------------------|---|
| _____ | 1. Hypothalamus                 | a. A treatment used to help treat sleep apnea   |
| _____ | 2. "Chase and Pace"             | b. A breathing disorder that occurs during sleep in which air is not able to enter the lungs                        |
| _____ | 3. Gastroparesis                | c. The area of the brain that does not work properly in persons with PWS  |
| _____ | 4. Obstructive sleep apnea      | d. A strategy that encourages slowing the rate of eating and helping to add moisture and facilitate safe swallowing |
| _____ | 5. CPAP                         | e. Slow emptying of the stomach   |
| _____ | 6. "Squatty Potty"              | f. Signs of an infection  |
| _____ | 7. Calcium/Vitamin D            | g. A resource that contains information about health issues in persons with PWS                                     |
| _____ | 8. Redness, warmth and drainage | h. A medical condition that results when a person drinks too many liquids at one time                               |
| _____ | 9. Medical Alert booklet        | i. Vitamins used to help treat fragile bones  |
| _____ | 10. Water Intoxication          | j. A stool used to help position the colon to help a person have a BM.  |

#### True or False (Circle T if True or F if False)

T or F	11. You are an important part of the team caring for the person with PWS. It is often necessary for you to help educate and advocate with health care professionals.
T or F	12. When a person with PWS eats, the message of fullness does not reach their brain.
T or F	13. If a person with PWS vomits, you should encourage rest but no further action is needed.
T or F	14. It is not necessary for food security to take place when a person with PWS is hospitalized.
T or F	15. Food security involves many different strategies that can be lifesaving to the person with PWS.
T or F	16. A low production of saliva and eating very fast are 2 factors that put a person with PWS at risk for choking.
T or F	17. Binge eating and/or eating large quantities of food at one time is not a big deal as long as the food is low in calories.
T or F	18. It is important for all injuries and complaints of pain to be taken seriously in a person with PWS.
T or F	19. Many persons with PWS may not have a fever when they are sick.
T or F	20. Drinking too many liquids at one time is not a problem as long as it is water or a low-calorie beverage.

**True or False (Circle T if True or F if False)**

T or F	21. If a person with PWS has a bowel movement every day, there is no concern that he/she may have a problem with constipation.
T or F	22. If it is a very warm day, it is important to encourage the person with PWS to dress appropriately for the weather and limit their time in warm environments.
T or F	23. Having low bone density (fragile bones) is a problem that often requires people with PWS to take medications and/or hormones to help their bones get stronger.
T or F	24. It is rare for persons with PWS to experience emotional and behavioral challenges.
T or F	25. Keeping hands busy and limiting time alone are 2 things you can do to help persons with PWS who pick at their skin.
T or F	26. The only reason people with PWS gain weight easily is because they have a low metabolism.
T or F	27. Choking is not a common problem seen in persons with PWS.
T or F	28. Going for a walk, even a short one, after eating assists with digestion, helps move food from the stomach and into the intestine as well as prevents constipation.
T or F	29. A person with PWS often has a lower than normal body temperature.
T or F	30. It is not a problem to administer medication for the treatment of diarrhea for persons with PWS.

**Multiple Choice Questions (Circle the correct answer)**

31. Tony has just fainted and you must take him to the Emergency Department at the local hospital. What resources can you use and/or share with medical staff that will be caring for him?
- PWSA (USA) Medical Alert booklet or handout
  - PWSA (USA) handouts
  - PWSA phone app
  - All of the above
32. Sara slipped and fell down a few stairs. She says she twisted her ankle but she reports that it doesn't hurt too much. What things should you do?
- Nothing – since she says that it doesn't really hurt.
  - Check it to make sure it is not deformed or swollen.
  - Apply ice and elevate. If she is unable to walk normally on it, take her to a clinic or Urgent Care for evaluation.
  - B and C
33. What are serious health problems that a person with PWS who is very overweight may experience?
- Headaches, diarrhea and diabetes.
  - Heart and circulation problems, breathing difficulties and diabetes.
  - Vision problems, breathing difficulties and stomach problems.
  - There are no serious health problems.
34. Things you can do to help with food security include:
- Use locks and/or alarms.
  - Plan, structure and prepare all meals.
  - Make sure that a plan is in place if an outing impacts snack or meal time.
  - All of the above.

35. You are responsible for preparing meals for the residents you support. What can you do to help prevent or manage the problem of slow stomach emptying?
- Limit the amount of liquids you use to cook and prepare the food.
  - Provide lots of salads and raw vegetables (high fiber foods).
  - Soften foods by cooking vegetables and serving fruits in natural juices and/or sauces.
  - Make sure to serve lots of different food items at one meal.
36. A tool you can use to help you judge if someone's stool (poop) is showing signs of constipation is:
- Bristol chart
  - Snellen eye chart
  - There are no tools to help with this task.
37. It is very cold outside and Jon, one of the residents you support, is refusing to wear a warm coat and hat. What should you do?
- Nothing – he will just have to go without appropriate warm clothing.
  - Attempt to explain how important it is to dress warm in a calm voice.
  - Don't argue with the person and bring a hat and warmer clothes with you. (Maybe he will put it on later).
  - B and C
38. You bring Adam, a young man with PWS, to the Emergency Department because he vomited after eating his lunch. He has been diagnosed with appendicitis and is being prepared for surgery. What can you do to help inform the anesthesiologist about PWS?
- Tell the doctor that Adam has PWS and assume that the doctor will know about his sensitivity to anesthesia.
  - Use the PWSA (USA) phone app and locate the handout on "Anesthesia and PWS" and share it with the doctor. Emphasize the seriousness of this concern.
  - Don't say anything. You are sure everything will be okay.
  - Talk to the nurse who is caring for Adam and hopefully she will inform the doctor.
39. What are some of the medications that persons with PWS may be extra sensitive to?
- Pain medications – especially narcotics
  - Anti-diarrheal medication
  - Sedative medications including anesthesia
  - All of the above
40. Beth has an extreme problem with skin picking. What are some things you can do to help her with this problem?
- Nothing – there are no cures for this problem.
  - Provide her with activities like a stress ball or lotion to help her keep her hands busy.
  - Cover any open sores with Band-Aids and/or clothing that will act as a barrier and help prevent her from picking.
  - B and C
41. Brian fell last night while on a walk. He complained of minor pain but there was no evidence of swelling. Today he awakes and his right ankle is very swollen and he states it continues to hurt. What should you do?
- Put ice on it for 20 minutes and send him to work. Have the evening staff check it again when he gets home.
  - Keep an eye on it and if it is still hurting him later, call and make an appointment for him to see his doctor.
  - Since the problem has gotten worse, make arrangements for Brian to be seen by his health care provider or taken to an Urgent Care today.
  - Nothing – you are sure it is a sprain and it will get better in a few days.
42. Kate has problems with her feet and ankles becoming very swollen – especially when she sits for a long time. What can you do to help?
- Check her temperature twice a month.
  - Make sure she gets up every 1-2 hours for a short walk to help with circulation.
  - Use circulation stockings if her health care professional has ordered them for her.
  - B and C

43. Hailey has had a persistent cough and congestion for 3 days now and she seems to be getting worse. Today she seems to be short of breath and more tired. She states she feels so bad that she does not want to go to work today (which is very unusual for her). What should you do?
- a. Let her stay home from work today and allow her to sleep all day.
  - b. Have her evaluated by a health care professional today to make sure she is not developing pneumonia.
  - c. Wait another day to see if she gets better. It often takes time for a cold to improve.
  - d. Give her some cough medicine and send her to work.
44. Laura eats her food very fast. You are providing oversight during lunch today. What are some things you can try to help her eat slower?
- a. Encourage her to put her fork down between each bite.
  - b. Have her take a sip of water between each bite.
  - c. Don't serve her any food unless she promises not to eat too fast.
  - d. A and B
45. Joe has been complaining of a sore throat for a few days. His temperature is 98.9 (His normal baseline temperature is 97.4). You take him to the Urgent Care and the doctor reports that Joe's throat is a little red but since he is not running a fever, it is probably a virus and he will be fine in a few days. What more can you do to make sure Joe's doctor has a good understanding of PWS?
- a. Show the doctor the Medical Alert booklet from PWSA (USA) that outlines the body temperature abnormalities and the lack of fever that is often seen in persons with PWS.
  - b. Nothing. The doctor knows best and you do not need to educate him/her.
  - c. Ask if Joe could have a throat culture since he often does not run a fever when he is ill.
  - d. A and C