

Find Your Voice:

Advocating for PWS Health Care Webinar Series



Health Education and Advocacy for Parents and Caregivers

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Session 2

OBJECTIVES



**Grow confidence in
health advocacy**

**Identify ways to
find information**

**How to come
prepared**

**Grow confidence
in more common
urgent health
issues**

PRIMARY ADVOCATES



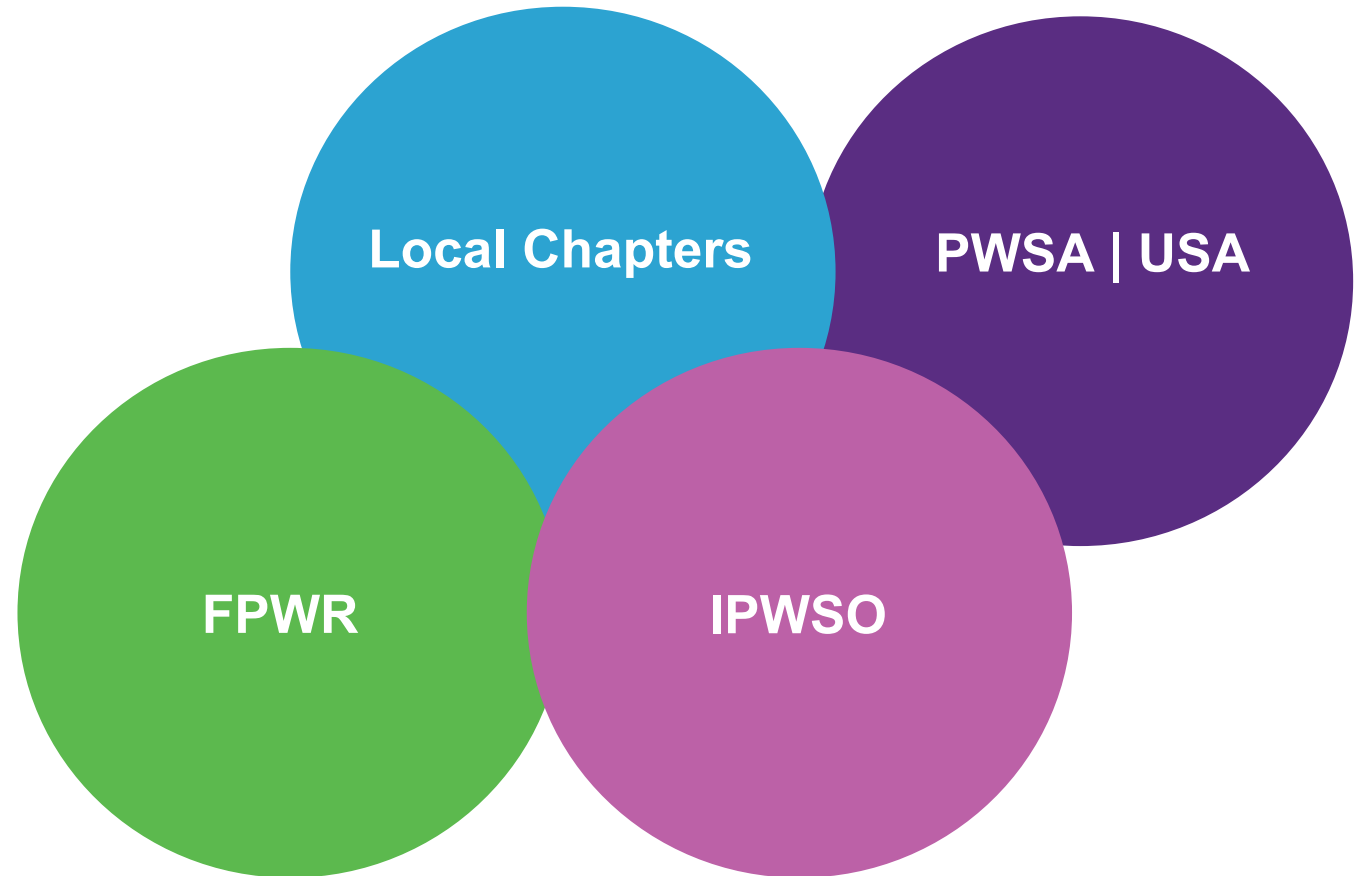
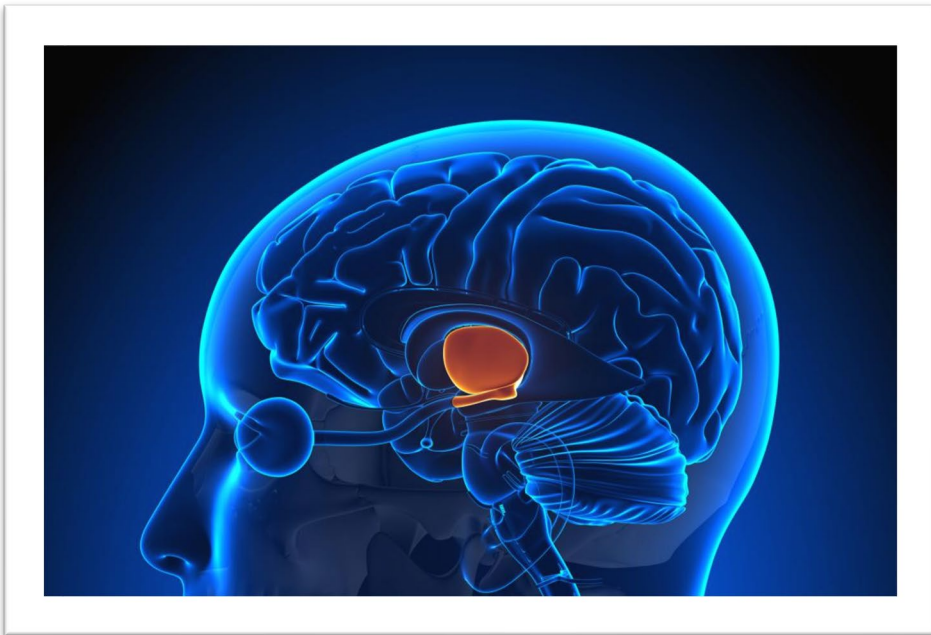
Parents and Caregivers are the primary advocates throughout the life span.

The first step in confidently advocating is becoming informed and educated.

STAYING ENGAGED AND INFORMED



There are numerous ways to stay informed



TOPICS FOR DISCUSSION



GI problems

Fragile bones

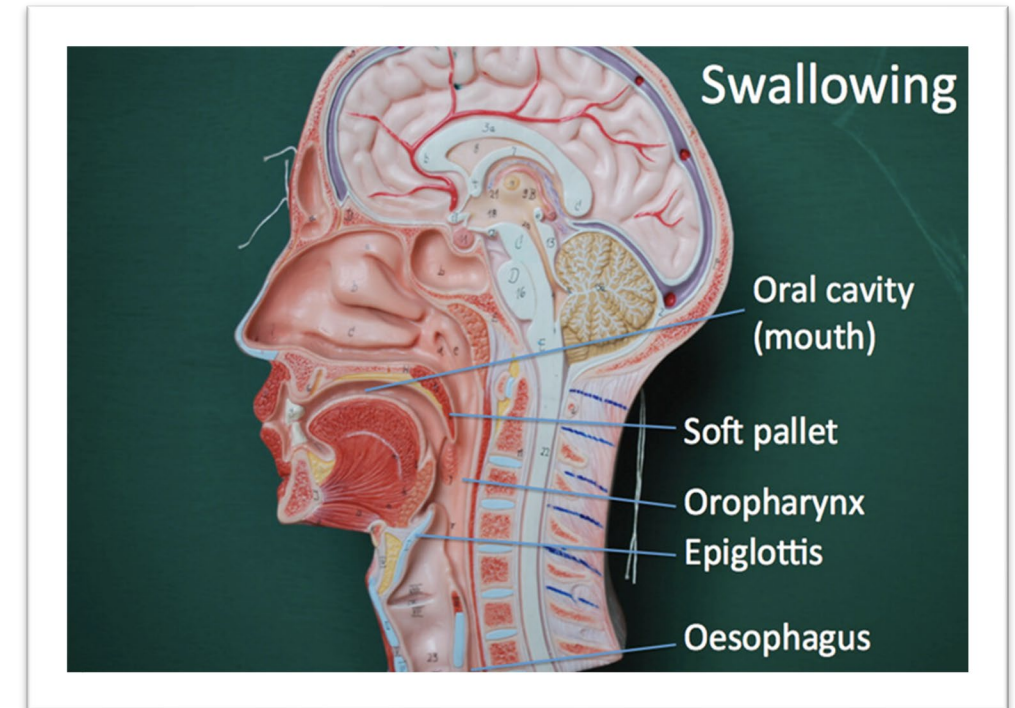
**Food seeking
and weight
management**

Skin picking



Swallowing difficulties

- It is very common for infants to have problems swallowing and require specialized feeding techniques. Following with speech and or a feeding specialist is important. Requesting evaluations may be necessary
- Research shows many children and adults have swallowing difficulties, which puts them at risk for choking and aspiration. This can be subtle or “silent”
- Many people with PWS eat very quickly and do not chew adequately, increasing risk for choking





**Heimlich
maneuver
training**

**Supervised
meals**

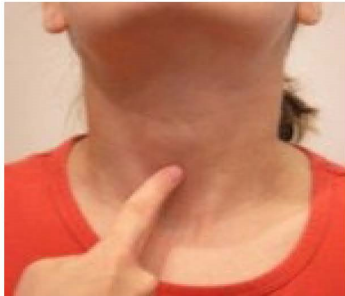
**Treatment of
reflux**

**Advocacy for
swallow
eval/study**

PACE AND CHASE



Why is “Pace and Chase” important?



Sometimes when I eat, food gets stuck in my throat, but I don't feel it.



I take a drink after two bites so that all the food goes to my belly. This is called “Pace and Chase.”



People remind me to take drinks. They care about me and want me to be safe.



I ask for water when my first drink is empty.



At the end of my meal, I drink my “flush” to make sure there is no food in my throat.

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TIPS FOR SLOWING DOWN

1

Put utensils
down between
bites

2

Serve smaller
portions

3

Cut up food
prior to serving

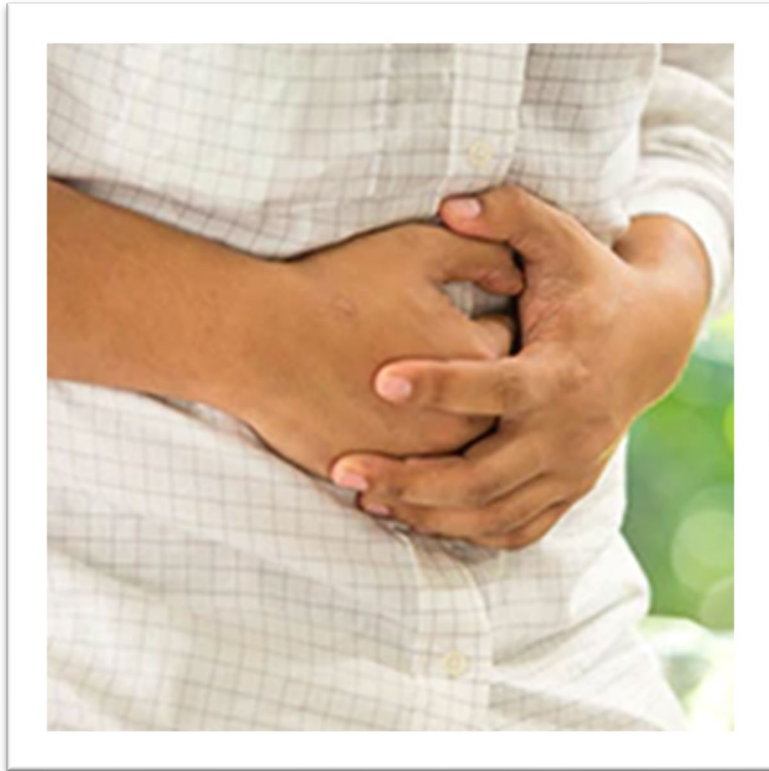
4

Provide food
with moisture
(soups and
sauces)

STOMACH PROBLEMS



Many individuals have slow or delayed stomach emptying



Bloating or distention of the stomach

Vomiting - people with PWS rarely vomit (please see GI Algorithm)

Remember that PWS individuals have a high pain threshold and may not report stomach pain

STOMACH PROBLEMS



Things that may make this worse



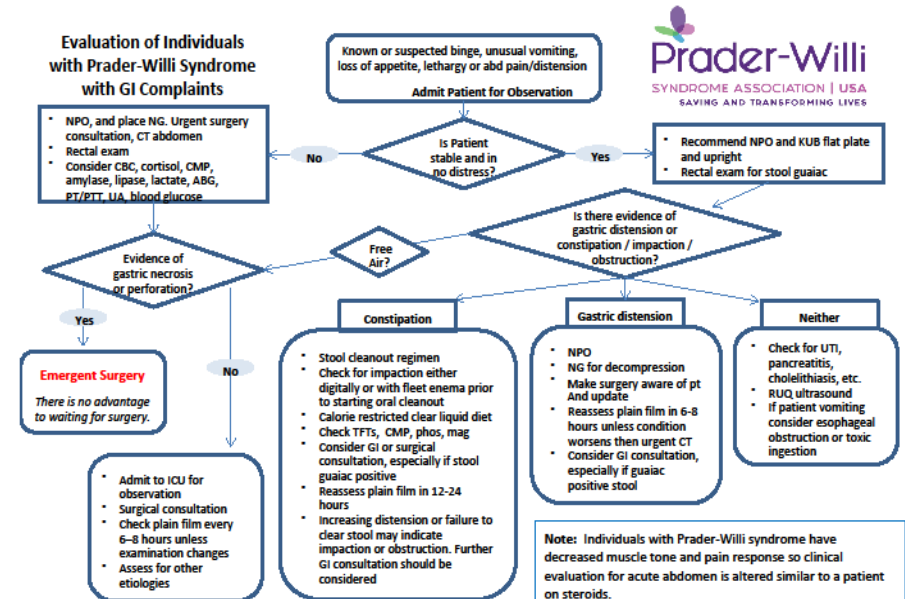
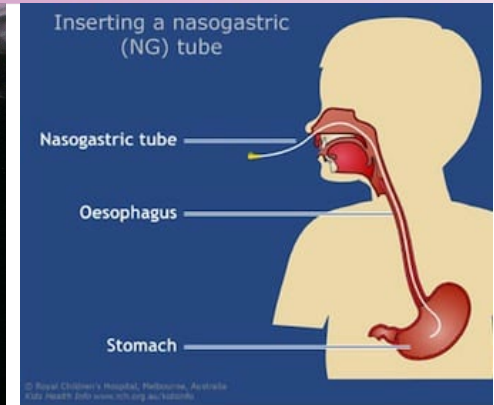
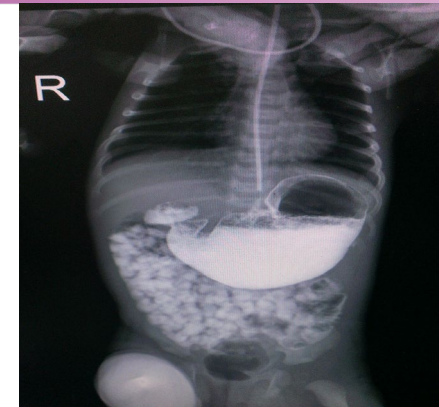
**High fiber and
fatty foods**

**Overeating/
binge eating**

ADVOCACY TIPS



- Medical Alert Booklet
- Doing an x-ray or other imaging will help them see if the stomach is distended
- A nasogastric tube (NG) may be needed to decompress the stomach
- They may not be able to eat or drink
- Ask that a gastroenterologist be consulted



Credits: James Loker, M.D., Pediatric Cardiologist • Ann Scheimann, M.D., M.B.A., Gastroenterologist • PWSA (USA) Clinical Advisory Board
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CONSTIPATION

Common causes

	Type 1	Separate hard lumps	SEVERE CONSTIPATION
	Type 2	Lumpy and sausage like	MILD CONSTIPATION
	Type 3	A sausage shape with cracks in the surface	NORMAL
	Type 4	Like a smooth, soft sausage or snake	NORMAL
	Type 5	Soft blobs with clear-cut edges	LACKING FIBRE
	Type 6	Mushy consistency with ragged edges	MILD DIARRHEA
	Type 7	Liquid consistency with no solid pieces	SEVERE DIARRHEA

**Decreased
physical
activity**

**Poor fluid
intake**

**Poor muscle
tone and slow
motility**

**Not enough
fruits and
vegetables**

Medications

ADVOCACY TIPS



**Discuss with
PCP**

**If persisting -
ask for a GI
referral**

**Encourage
activity**

Keep records

Offer fluids

**Well balanced
diet**

**Try a squatty
potty**

**Medications as
prescribed**

FRAGILE BONES



**Low bone
density**

**Low vitamin and
mineral levels**

**Hormone
deficiencies**

**Decreased
weight bearing
activity**



Fractures occur easily



Poor body/special awareness



Decreased pain may lead to fractures not being identified early

ADVOCACY TIPS



**Monitor after a fall
and seek medical
care if needed**

**Request x-ray or
other testing if you
feel necessary**

**Get routine labs like
Ca, Vit D, DEXA
scans, hormone
levels**

SKIN PICKING

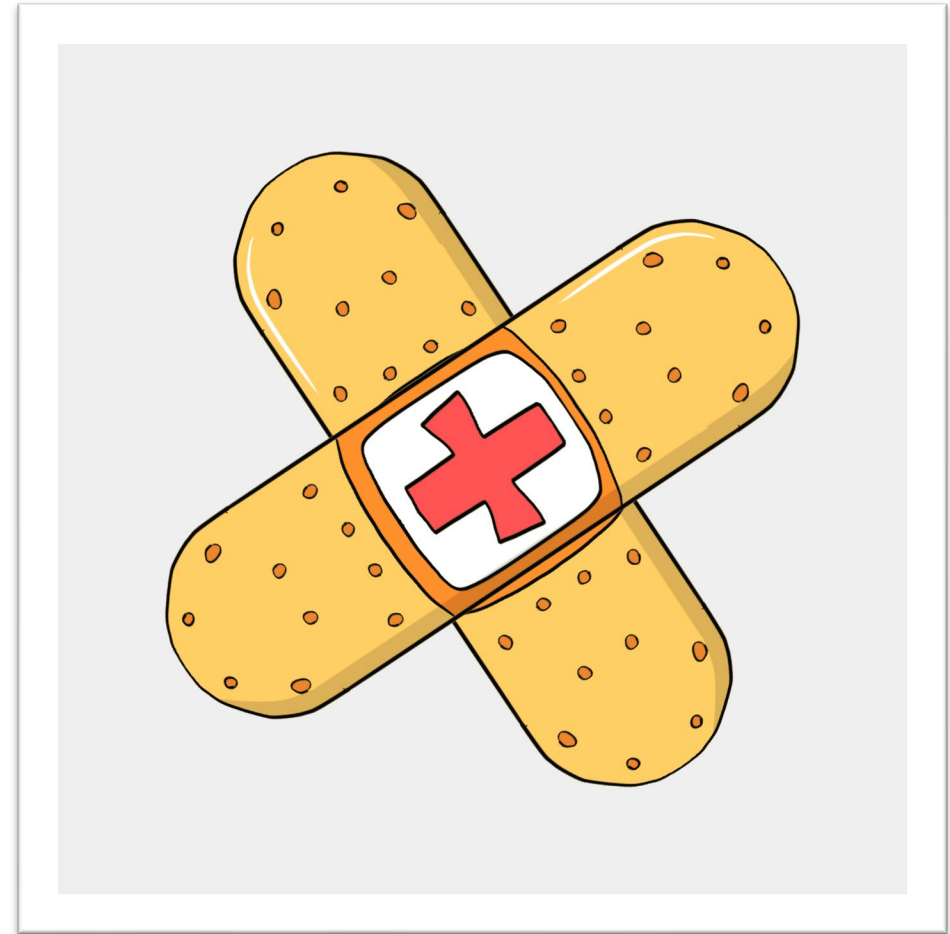


- Skin picking/rectal picking are common with PWS
- Preventing, treating, and managing can be very challenging
- Goals include prevention of infection and healing of open sores

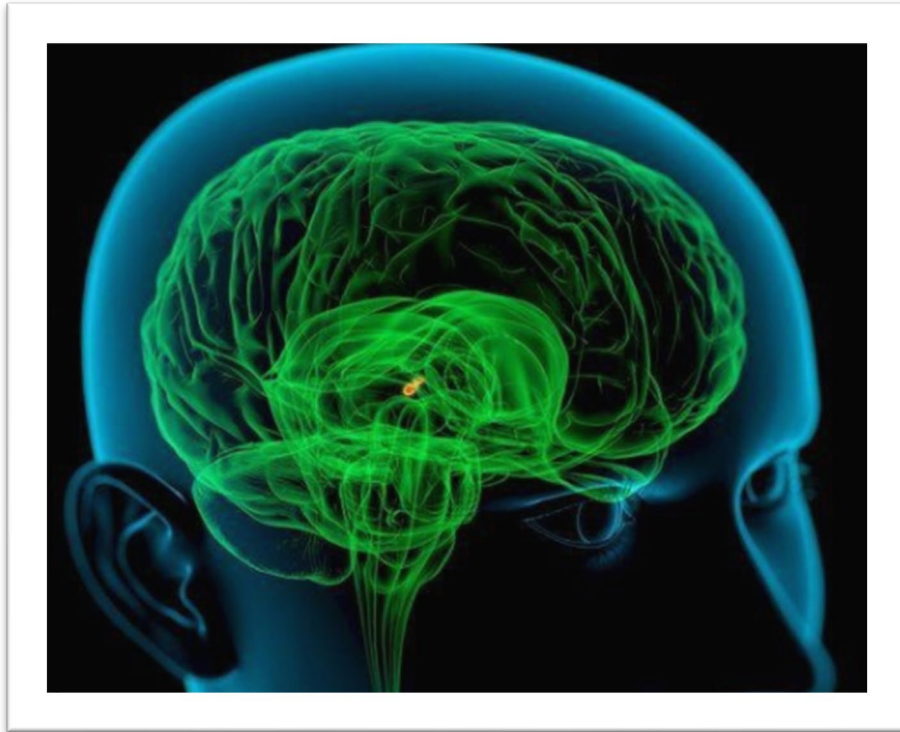
SKIN PICKING TIPS



- Regular sensory activities
- Keeping nails short
- Reward positive behavior relating to not picking or good skin hygiene
- Discuss medications like Naltrexone or N-Acetylcysteine, which have shown success in **SOME** individuals
- Keep hands busy



FOOD SEEKING AND WEIGHT MANAGEMENT



Nutritional phases

1A

1B

2A

2B

3



PWS is VERY complex



- **Hyperphagia** is defined as abnormally increased appetite of food frequently associated with injury to the hypothalamus
- This increased appetite we know includes a preoccupation of anything food-related can lead to increased 'anxiety' in our loved ones

WEIGHT GAIN



- People with PWS can gain weight on half of the calories of their peers
- Incorporating healthy food and exercise choices from the very beginning helps set up for long-term success
- There is no magic diet for everyone with PWS
- Growth hormone is helpful for lean muscle mass, which helps metabolism and burning calories
- Food security options may include locks on refrigerators, cabinets, and other sources of food
- Incorporating daily exercise will help with weight management



ADVOCACY TIPS



Food security at home/school/work

**Open communication with
family/friends/teachers/co-workers**

Stay informed of clinical trials

Seek help of dietitian

Medical Alert Booklet



Open Discussion

Q & A



RESOURCES

PWSA | USA Medical Alert booklet



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