



5TH  29th to 31st oct 2021
ASIAPACIFIC
Prader-Willi Syndrome Conference
One Voice Together

HEALTHY AGING in PWS

Susanne Blichfeldt.MD
Denmark

s.blichfeldt@dadlnet.dk



Aging. Many aspects.

- Body- Health
- Mental- Health.... being happy.
- Life situation: Where to live, to have support
- During the day: School, Job, Activities, Hobbies.
- Social life: Family and friends.
- Parents and family are important. Support.
- Who will take over ?

What we know today about PWS adults

- More and more adults are living healthy , happy with normal weight
- It is possible to have a long and healthy life with PWS
- Special support is always needed, together with realistic expectations
- Family, caregivers and health professionals must know PWS
- When diseases arise and specific help is needed.

And when you are old: PWS and all others

- Looking older
Less muscle power.
Balance can be weak. More weak than before (PWS)
- More risks for diseases.
- Need for help can change, when you are not able to do as before
- Mentally : you are the same... and have more experience
- But with PWS it can be difficult to explain the changes that come with age

Diseases in PWS

- Symptoms can be different with PWS compared to others
- The body often reacts differently than expected with diseases
- Examples: Less pain, no fever with infections is often seen
- People with PWS can perhaps not explain if they are ill
- Therefore, there is a risk that diseases can be overlooked
- Means : a risk that diseases are diagnosed late, or even too late

Symptoms and diseases in PWS

- Special symptoms can be seen in PWS with diseases in general
- Diseases because of overweight
- Diseases and symptoms related to special organs/areas in the body in PWS, can come with age, and are not always expected.
- Delayed diagnoses can cause early and unexpected death

Causes of deaths in adults with PWS

- Overweight
- Respiratory problems: Lung infections not treated
- Abdominal diseases: Infections and stomach rupture
- Accidents: Also in traffic
- Choking: Breathing is blocked by food in the throat.

The role of Hypothalamic Dysfunction

- Appetite : Always a desire to eat, risk of severe overweight
- Hormones: Growth hormone, sex hormones, thyroids: can be low
- Temperature: Severe infections without fever
- Sensations: Do not feel warm and cold as we do: need help to dress

- High pain threshold: Do not feel pain and do not complain :
therefore: abdominal diseases and fractures can be overlooked

- Sleep regulation

The body and PWS

- Lean body mass is small in PWS. (also with GH)

- Lean body mass:

The body without fat: muscles, bones ,organs:

- The fat on the body in PWS:

The fat mass is "big", even with normal weight:

- Muscles and bones can become even "smaller" with age, with inactivity
- The muscles are using the calories eaten
- Are the muscles small and inactive, calory needs are very low (PWS)

How much to eat, changes with age?

- If a person with PWS eats the same as others, the person puts on weight.

- Great variations:

Having a daily/weekly weight will tell. Weight gain: what is eaten ?

- With overweight:

Calory needs are low: 6-8 kcal/ cm height or less (depends how much you move)

Because of small lean body mass and or inactivity: do not move so much

- A risk:

Calory needs go down with age (with inactivity)

Medication and anaesthesia

Medication:

Start with low or ½ dose to avoid side effects

Because of small lean body mass, (the medication goes to the fat free part of the body first) Be careful with medication that can give sedation

Weight does not go up because of a medication itself. It can perhaps change appetite
If you eat more and move less weight goes up. Take care of the food.

Anaesthesia:

Waking up can be delayed

Apnea can arise after anaesthesia

Advice: stay overnight after an operation in hospital, monitor respiration

Overweight can cause diseases

- **Overweight gives with age risks of**
- Hypertension and heart failure
- Diabetes 2: can harm kidneys and eyes
- Joint problems
- Poor circulation: Edema and leg ulcers
- Sleep apnea
- Restricted ventilation(lung problems)
- Movements become difficult, move less, gain easily weight
- Hygiene can be difficult.

Gastrointestinal problems can worsen with age

- Reflux: acid from stomach runs back to the mouth. Harms the teeth.
- Teeth: can be worn down. Need treatment.
- Teeth infections can come, and be overlooked (no pain etc)

- Distended stomach: after eating, with diseases, with constipation
- Cannot vomit. **Only symptom can be: do not want to eat.**

- Constipation: common, more common with age? often not known

Kidney and bladder

- Most adults have urinary control
- Incontinence: can arise with obesity
- Many do not empty bladder, sensation and urge is not normal
- Urinary infections: With poor hygiene and overweight.
- To recommend: for some
- Toilet visits at fixed hours: so that bladder is emptied.
- Many need more time than usual

Bones, legs and circulation

- Osteoporoses is common in PWS. Hormone deficiency can worsen
- Daily vitamin D is needed.

- Legs:
- Poor circulation can cause edema , worsen with age in some
- Treatment: moving, diuretics does not help

- Leg ulcers can be infected. Especially with edema and skin picking

- Need of special soles in shoes? Look at feet. Malformations ?

The back and the spine in PWS

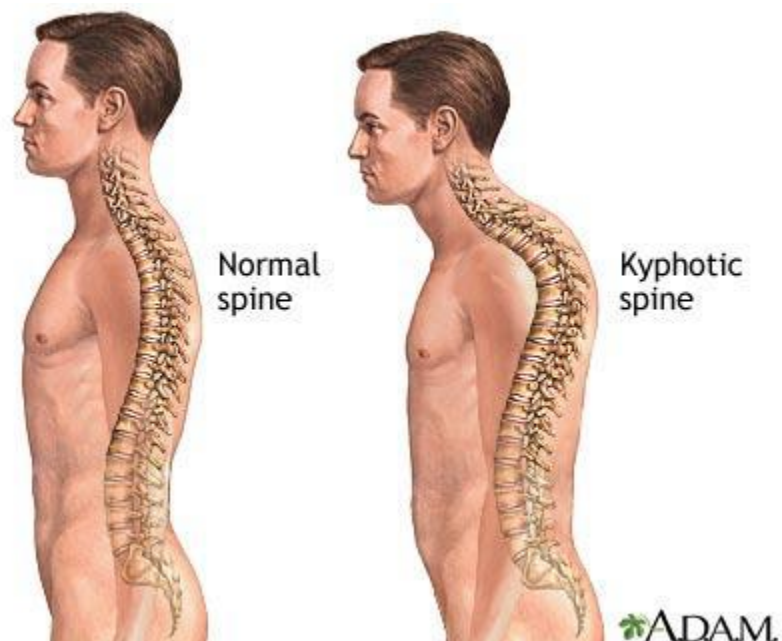
- SCOLIOSES: Spine has a S form
- Not rare, can be severe and restrict ventilation, worsen with age, give pain
- Treatment: Consider operation

- KYPHOSES: Overbending of spine. Can worsen with age. Arise because of muscular hypotonia
- ***Dayly exercises***, to strengthen the back, ***can help to avoid permanent fixed overbending.***

- ***Also in aging adults. Not too late to start!!***

Kyphoses und Scoliose

- Kyphoses



- Scolioses



Also remember

Eyes:

- Vision can become weaker with age: check every 2-3 years

Hearing:

- Remember to check all with PWS, and after age 50 ,
- Every 3-4 years or with symptoms, not all can tell that hearing is bad

Behavior and Psychiatry

- Behavior is special in PWS.
- Need of security and routines. To be prepared for changes.
- Anxiety is often seen if things and food are insecure, also in adults

- Psychiatric diseases can arise:
- Depression and psychoses, can be treated with medication
- Correct diagnoses is important. The person has changed
- Dementia is rare
- Read: www.IPWSO.org/index/ >>> medical professionals >> mental health

Mental health and PWS

- Anything changed ?
- Remember each year to write a „story of the year“
- DIARY OF LIFE
- So that staff also know how was life „before“
- Also to be able to talk about things that happened before

How to know: Is he or she ill ?

- You cannot rely on temperature or fever
- You cannot rely on complaints about pain
- You must look at the general well being:
- Something changed: Appetite ? Behavior?
- Is it a physical or a mental problem?
- Something in the environment ?

- If he/she **does not want to eat or vomit** : something is very wrong

- Consider if problems are caused by new medication , especially psychotropics?

Make a PWS ID card, credit card size

In your own language and in English

- TEXT:

- The holder of this card has Prader-Willi Syndrome (PWS)

- These symptoms require immediate medical attention:

- Loss of appetite, vomiting or bloated abdomen (consider ileus)

- Withdrawn or obvious change of behaviour (consider psychosis)

- Complaints of new pain

- Reduced function of any part of the body (consider bone fracture)

ID card: Page 2:

- Notice that a person with PWS can have:
 - Difficulties in explaining own symptoms
 - No fever in connection with infection
 - Increased pain threshold
 - No ability to vomit
 - Increased risk of apnea related to use of sedatives and tranquilizers
- Further information: www.ipwso.org and your national association
- Contact: address for the hospital where you are known

How to keep adults healthy and fit. It is simple and cheap.

Move:

- Walk every day, run if possible!! : good for bones!

Food:

- Only eat at meals. Only one plate. Have a plan

Hygiene:

- Have good routines for every day

Happiness:

- Do things you like. Have success. Responsibility for what you can. Self esteem

How to move more

Have a plan:

- After each meal: walk and talk: best ½ hour, take the same tour, is OK
- Have favorite places to go to: visit animals, visit museums
- Take stairs, no lift
- Go dancing:
- Have a plan at home: music and dance
- Swimming, fitness, whatever possible

- Be aware that he/she with PWS is not sitting most of the day.

Food and PWS

- Calory needs can be very low, if you move very little
- Ask the doctor and dietist if possible (they must know PWS)
- Reduce fat : minimum of oil, no butter, no sugar. One fruit per day, or ½ banana.
- Vegetables : boiled or raw in small pieces
- Avoid oil in sauces, use vegetables and spices.
- Drink water !
- Remember vitamin D

To be happy, and no stress: A plan for the day

- Security is the most important in PWS. To feel secure.
- About food: to know when you have meals.
- About what to do and when. And together with whom
- Have day plans: Written and with pictures
- Set the time:
- So you know when you start and when you finish an activity
- Occupation with what you like to do.
- Do not skip an activity because of age, but reduce hours, if needed

CONCLUSION : Symptoms in PWS

In acute situations: the unexpected reactions and symptoms

- Temperature and sensation of pain
- Overfilled or distended stomach , do not vomit.

Daily care and possible symptoms to know, not related to weight:

- Eye and vision, teeth, skin (skinpicking), poor chewing , scolioses und kyphoses, Intestine and bladder function. Tiredness, apnea, too much eating.
- Side effects of medication

Symptoms because of overweight.

- Heart and lung function, edema, incontinence, leg ulcers, restricted motor function, sleepiness

Every year at the doctor

Check:

- Heart, Lungs
- Circulation
- Joints
- Teeth and skin

- Blood tests:
- Hormones, vitamin D
- Cholesterol, lipids
- Diabetes? Blood sugar tests

Write a book.

A life story book.

Print photos.

Write to remember

So nothing is forgotten about the wonderful person with Prder-Willi Syndrome

Conclusion: The importance with aging

- Being healthy and happy:
 - Moving
 - Healthy food
 - Normal weight
 - Being happy
 - Much support, success and love
-
- And that we all accept PWS and the needs there are with PWS

From age 3 to 39

Happy
with
support



THANK YOU from Denmark

