

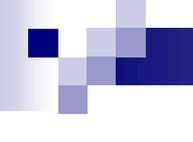


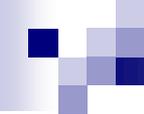
Nutrition in Prader-Willi syndrome challenges beyond obesity

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Dysphagia assessment and diet management in PWS

- Infants with PWS demonstrate feeding difficulties related to the neuromuscular finding of hypotonia.
- They have a high risk of feeding and swallowing dysfunction.
- Deaths in infants with PWS are most likely to be related to respiratory failure, aspiration, infection, and choking.

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- Stevenson DA, *et al*, (2007) Choking incidence of 34% and 8% mortality from reviewing deaths of 152 individuals with PWS
 - Children with PWS have an increased risk of respiratory morbidity, and food aspiration increase the risk.
 - Swallow dysfunction and aspiration risk change over time.
 - Swallowing assessment is a must in nutritional approach for PWS.

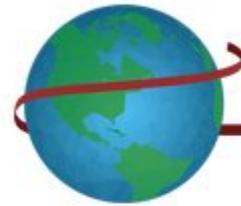
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- Children and adults with PWS can have important physiologic signs of dysphagia and risk factors for choking and aspiration
 - Gross R, *et al*, (2016) proved that swallowing impairments are physiological, subclinical and not only related to neurobehavioral factors
 - Subclinical dysphagia emerge from the lack of typical clinical signs of dysphagia in the presence of direct fluoroscopic evidence indicates that the dysphagia, and it cannot be detected without instrumentation.

The need for an instrumental swallow test in PWS

- It is recommended that persons with PWS receive video fluoroscopic swallowing evaluations, particularly those who have a history of regurgitation, choking or pulmonary infection.
- swallowing evaluation should be conducted with the patient in their natural seated position, use solid food items under videofluoroscopy and even fiber optic endoscopic evaluation of swallowing

Multidisciplinary approach

- Team for swallow test – diagnosis staging the dysphagia level
- Determine appropriate nutritional interventions:
 - including diet/meal pattern,
 - nutritional supplements,
 - assistive devices as needed,
 - food texture and liquid consistency modifications,
 - positioning and route of feeding (including enteral and parenteral feeding regimes)



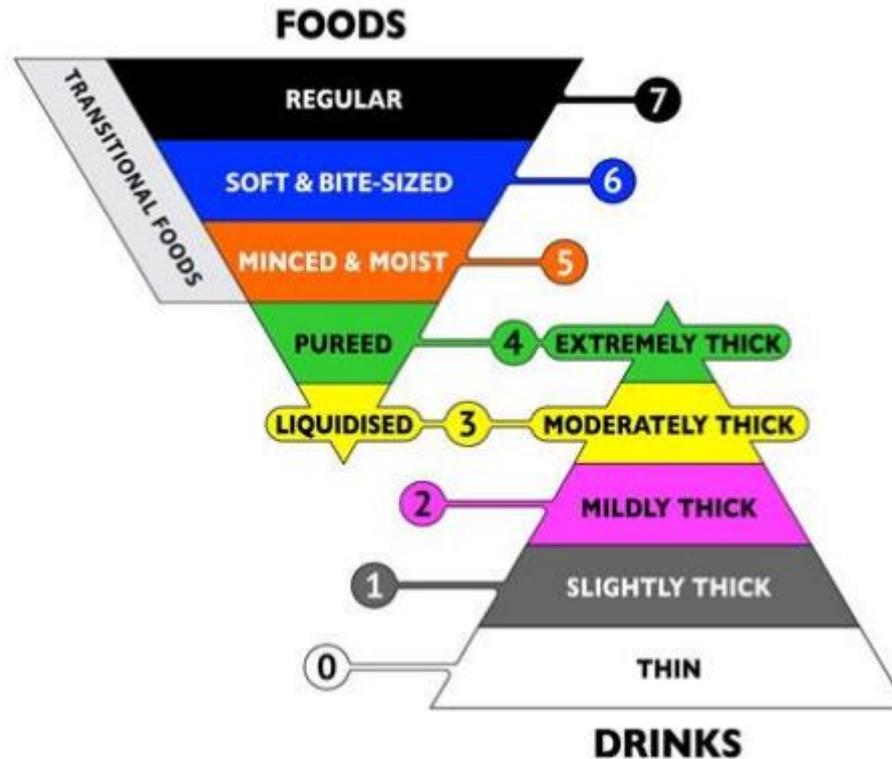
IDDSI

International Dysphagia Diet Standardisation Initiative

A global initiative to improve the lives of over 590 million people worldwide living with dysphagia

- a global terminology that will ‘work’ for all cultures and that will be accompanied by practical and valid measurement techniques that will facilitate use by persons with dysphagia, caregivers, clinicians, food service professionals and industry partners.
- a standardized way of naming and describing texture modified foods and thickened liquids for people with dysphagia across the lifespan

International Dysphagia Diet Framework implementation in Romania 2018



Mapping to IDDSI - Foods

Current NDD Food Textures

	Regular
	Dysphagia Advanced ✓
	Dysphagia Mechanically Altered ✓
	Dysphagia Pureed ✓
	



	7 Regular
	6 Soft & Bite-Sized
	5 Minced & Moist
	4 Pureed
	3 Liquidised

Transitional Foods





Thumb nail blanching to white



Sample squashes and fractures, and does not return to its original shape when pressure is released

The International Dysphagia Diet Standardisation Initiative 2016 @<http://iddsi.org/framework/>.

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- It is possible that instrumental swallowing assessment and implementation of a matching texture standardised diet with a multidisciplinary team, will reduce the risk of death in people with PWS.



Thank you!

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