

How to fit intake of cysteamine with other medication, food, drinks and tube feeding?

A Challenge...

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Case report – Girl 10y with cystinosis

14 months old; Diagnosis of Cystinosis

- Failure to thrive
- Renal Fanconi syndrome
- Start Cystagon
- Supplements for Fanconi syndrome

9 years old;

- Switched to Procysbi





This is a daily dosage of medication for Denice Flerchinger's teenage daughter, who has cystinosis.



	7.15u	Tss 7.15u en 8.15u niet eten	Vanaf 8u15 ontbijt. Na ontbijt rest medicatie	13u = na middagmaag	15u30	Voor 18.15u eten,	19u15	Vanaf 20.15 terug eten	Voor slapengaan lkr (na 23u)
D-cure		N I E T S E T E N	5 druppels			N I E T S E T E N		N I E T S E T E N	
Fosfaatsiroop (Potio van Jolie)			7ml	7 ml	7 ml		7 ml		
Natriumcitraat 500mg			1	1			1		
Kaliumcitraat 1000mg			3	3	3		3		3
Kaliumcitraat 500mg			1	1			1		
Procysbi	10 (2x5/lepel)						10 (2x5/lepel)		
Lisinopril			0,6mg				0,6mg		
Cystadrops (oogdruppels)			1 druppel/oog	1 druppel/oog	1 druppel/oog		1 druppel/oog		
Groeihormoon (prikpen) FRIGO							1,0 via prikpen		

-Cysteamine **7.15h + 19.15h**

-1h before and after intake of cysteamine: fasting

-overload of medication at different times



Cysteamine in combination with medication and food -> struggles

- Nausea in the morning (GI disturbances , oral motor dysfunction)
- 1h before and after intake of cysteamine: fasting
 - No/late breakfast
 - No/late snack in the evening
 - Lack of calorie intake
- Hard to combine with other medication
- Lack of sleep
- Typical smell
- Social life (child- teenager –adult)
- Practical use of cysteamine

Need for individual approach!



Cysteamine and food intake – what's possible?

- Cysteamine bitartrate delayed-release capsules (Procybi®): coating
 - Bypass dissolution in the acidic environment of the stomach
 - Allow dissolution in the alkaline environment of the small intestine
- Selection of compatible foods:
 - Applesauce, berry jelly, fruit juices (except grapefruit)
- Extra?
 - $\text{pH} < 5,5$:Yoghurts, (pureed) fruits, yoghurt based smoothies, water, soda's
Other?
 - $\text{pH} > 5,5$: Baby formula, combined meals, plain milk, tube feeding, ...



The effect of food and liquid pH on the integrity of enteric-coated beads from cysteamine bitartrate delayed-release capsules (Drug design, Development and Therapy 2018:12)



Practical use of cysteamine

ORAL

- Capsules of cysteamine in combination with drink.
- Opened capsules in a drink (stir).
- Opened capsules in pureed fruits of yoghurt.

TUBE

- Opened capsules in a drink (stir). -> syringe
- Opened capsules in pureed fruits of yoghurt. -> syringe
- Pay attention: Rinse to take up all the medication and flush the tube with water



Cysteamine and food /medication intake – Your experience?

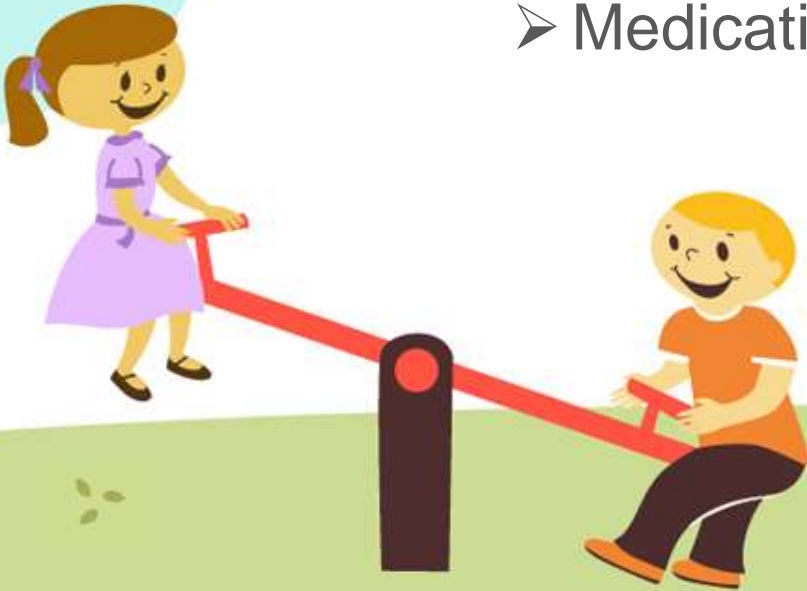
- Time of intake?
 - 1h before- after?
 - Strict (6-12h)?
- Combination with food?
 - PH < 5,5?
- Combination with medication?
- Administration by tube?
- Combination with tube feeding?

Need for individual approach!



Transition from tube to oral feeding

- Individual approach
- Demands changes
 - Plan feeding schedule
 - Medication intake moments





Cystinosis
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