



Digital Consultation: “Meet the expert” about nutrition and gastrointestinal problems in PCH2

Q&A

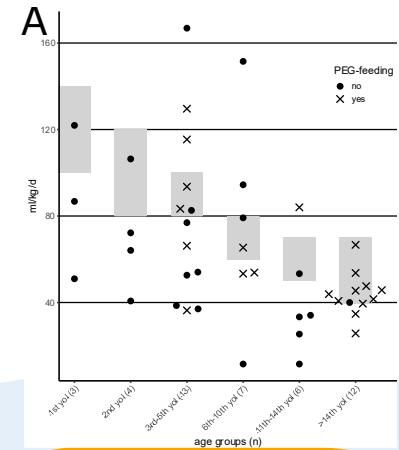
Disclaimer

The PCH2 treatment options listed here are based on data from the 2023 Natural History Study on PCH2 and personal experience from parents. The answers make no claim to be exhaustive and represent neither a specific recommendation nor an endorsement of the drugs or PCH2 treatment options mentioned. It is a compilation of measures that have been tried in the past and is intended for informative purposes only. PCH2cure assumes no liability in this respect.

Q&A session

Which foods slow down bowel activity?

- This can vary from person to person.
- Large quantities of cow's milk should be avoided, as they often lead to constipation.
- Low fluid intake: a common problem in children with severe multiple disabilities.
- High-fat foods.
- Foods with a high tannin content (unripe bananas, black tea).
- Foods with a high sugar content.
- High saturated fat content (especially animal products).



Q&A session

Can food in puree form be digested faster than chunky food (our daughter eats pasta and bread, for example), or is there no difference?

- The smaller the food is before it is eaten, the faster it can be digested.
- The “speed” of digestion varies greatly from person to person and depends heavily on the food.
- It depends on how the child is “feeling”.

Q&A session

Several small meals or a few large ones—what is the medical opinion on this for our children?

- In cases of recurrent vomiting and severe reflux, generally speaking, many small portions are better, BUT
- it must suit you, your child, and your everyday routine. In addition, the stomach needs a certain amount of food to grow.
- Caloric intake should be based on the age-appropriate norm—if in doubt, consult a nutritionist or have a blood test done: restless children tend to have higher caloric requirements, while calm children tend to have slightly lower requirements.
- Rule of thumb: there should be at least 2 hours between meals.

Q&A session

How can one estimate the amount of food consumed when eating solid foods?

- Use a 3-day food diary, which a nutritionist can use to calculate whether your diet is balanced or whether certain elements are missing.

Q&A session

My child sometimes eats solid food and occasionally swallows pieces. Is this a problem?

- Eating means quality of life → you must weigh up the risk of the child choking against improved quality of life and enjoyment of food.
- Soft foods can be fed, e.g., soft bread or pasta, but hard foods such as chips, cookies, or nuts should be avoided → children show through their behavior what they can and cannot eat.

Q&A session

Could digestive problems, such as constipation, be the reason why my child has adopted the “C-shape”?

- The exact cause of the C-shape in PCH2 is unknown.
- There is a hypothesis that pain, getting caught up in a (painful) situation or gastrointestinal problems promote the C-shape.
- It is best to try to prevent such situations from occurring in the first place.
- There is evidence that children with PCH2 adopt the C-shape less often as they get older.

Q&A session

Do febrile convulsions occur more frequently after taking Vomex (active ingredient: dimenhydrinate), or does Vomex lower the seizure threshold? What alternatives are there for nausea?

- Yes, in some cases Vomex lowers the seizure threshold.
- The drug of choice for children with neurological disorders is ondansetron → the advantage is that it does not lower the seizure threshold, causes less fatigue, and therefore does not disrupt the day-night rhythm.
- Alternatives for persistent symptoms: granisetron or aprepitant
- In general, the side effects of the drugs described are relatively easily predictable, but there is no data on their use in children.