

Supplement 2: Sample letter

Sample letter provided to the parents of a child with PKAN and severe dystonia

To Whom It May Concern:

___ is under my care in pediatric neurology for his dystonia due to Pantothenate Kinase-Associated Neurodegeneration (PKAN). Children with PKAN have severe dystonia and are at high risk for having status dystonicus/dystonic crisis (persistent, severe, life-threatening dystonia causing muscle breakdown, respiratory/cardiovascular compromise). Status dystonicus can be triggered by infection, constipation, pain, changes in medications such as missing regular dystonia medication, or anesthesia.

In order to avoid status dystonicus, please initiate the following important actions when ___ is coming into the hospital:

1. Call the hospitals' pediatric neurology consult service as soon as he arrives
2. It is imperative that he continue on his home medications without interruption of dosing unless doing so would be life-threatening. If he needs to be NPO, carefully consider the serious risks of his missing medications and determine whether he can still receive them. Even when he is otherwise NPO for medical/surgical reasons, he should receive his dystonia medications per g-tube unless doing so presents a specific documentable risk. He is typically on ___.
3. ___ should be allowed ___ to receive his CBD-THC (cannabis) oil which he receives at home as this can be very effective in stopping his dystonia. Other approaches to treating his dystonia include increasing ____.
4. If you are sending blood work, consider sending a serum CK to determine if he is having muscle breakdown from his dystonia.
- 5 Keep him cool with a fan or cooling blanket at all times to help manage his dystonia
6. He is at high risk for skin breakdown and fractures with severe dystonia - he should be on an air mattress and have padded rails on his bed with attention to blocking spaces that might entrap a limb.

Sincerely,