

Vanishing White Matter Patient Management Card

Recommendations for preventive measures and acute care settings

Introduction

- Vanishing White Matter is a rare genetic disease leading to progressive motor and cognitive dysfunction.
- Episodes of acute decline may occur. These episodes can be triggered by physical stressors, like fever, infections, head trauma and certain anesthetics.
- There is no cure for VWM, but there are best practices for clinical management.
- This leaflet contains key recommendations for preventive measures and acute care settings.

Do's

- Use ibuprofen in the case of fever, add paracetamol/acetaminophen if necessary
- Use antibiotics in the case of a (suspected) bacterial infection
- Propofol is preferred if, anesthesia is needed
- Treat epilepsy
- Follow the regular vaccination program, including Influenza and COVID-19 vaccinations
- Wear a helmet for activities with a risk of head trauma
- Apply standard behavioral management

Don'ts

- Contact sports
- Inhalation anesthetics

Recommendations for patients

Measures to prevent episodes of acute decline

- In the case of fever, use paracetamol/acetaminophen, ibuprofen or both to normalize body temperature.
 - In case of febrile infection, check with your physician if antibiotics are needed.
 - Follow the regular national vaccination program, including yearly Influenza vaccine and up-to-date vaccination against COVID-19, and give ibuprofen to avoid fever.
 - Wear a helmet for activities associated with enhanced risk of head trauma, such as bicycling.
 - Avoid contact sports.
 - Major surgical procedures carry extra risks for patients with VWM and benefits should therefore be carefully weighed before proceeding.
 - Standard behavioral management is safe, as psychological stress is not linked to acute decline.



Recommendations for physicians

Measures to prevent episodes of acute decline

- We recommend balancing the need for major surgical procedures against the risk of anesthesia and physical stress related to the procedure. PEG tube placement is a minor surgery. Other advices around surgical procedures are provided in the scientific publication mentioned in Read More.
- We suggest close monitoring after head trauma, surgical procedures and use of anesthetics.
- In the case of previous acute severe decline, we recommend to consider hospital admission with intensive care possibility when a similar stressor occurs.
- Consider avoiding drugs that activate the integrated stress response, especially inhalation anesthetics like sevoflurane. Use propofol instead.
- We recommend to start ibuprofen in the case of fever or vaccinations, aiming at normal temperature. Add paracetamol/acetaminophen if necessary.
- We recommend to maintain a good nutritional status.



Acute presentation management

- We recommend prompt investigations in the case of fever, with a low threshold for empiric antibiotics, if a bacterial cause is suspected.
- We recommend to consider hospital admission with intensive care availability in the case of acute neurological decline.
- A short course of corticosteroids can be considered in the case of acute severe neurological decline (e.g. with reduced level of consciousness, loss of walking), if there are no contraindications.

Questions? Please contact:
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Read more

The complete scientific article with all recommendations on diagnosis and management of VWM was published open-access (DOI: tba)

About us

This leaflet was established by VWM experts from the VWM consortium and patient advocates from the United Leukodystrophy Foundation (ULF), European Leukodystrophy Association (ELA), and Volwassenen, Kinderen en Stofwisselingsziekten (VKS).

Disclaimer

The information in this leaflet is carefully established based on scientific evidence and expert opinions. Future developments may lead to changes in these recommendations. Please check whether an updated version is available on <https://www.VWMconsortium.org/about-vwm/>