HEMIPLEGIC MIGRAINE ACTION PLAN

A hemiplegic migraine action plan contains guidelines on how to respond during an event. It includes health and medical information specific to the patient and guides the emergency department in the appropriate actions to take.

Patients with a history of hemiplegic migraine or with a *CACNA1A* variant known to be associated with hemiplegic migraine should have a written action plan, which includes health and medical information specific to the patient. Patients or caregivers and their neurologist should discuss and develop the plan together. It is important for the plan to be clear and concise because it may be utilized under very stressful situations (Hospital Emergency Room) and by individuals who may not know the patient.

A copy should be kept with the patient at all times. Many families have a hospital "go bag" which includes a laminated copy of their emergency plan and anything needed for an overnight hospital stay. Families may forward a copy of their emergency plan to their local hospital to be kept in the patient's file at all times.

What is Included in the Plan?

-Name of the patient, current age and diagnosis
-Emergency contact information
-Neurologist contact information
-Height and weight
-Listing of daily medications and doses
-List of drug allergies or medications the person should not take
-How to identify a hemiplegic migraine (if known, how the patient's HM presents)
-What rescue medications should be administered, when, and dosage. Can a second or third dose be given?
-Instructions for scans (MRI, CT) and labs

The plan should be signed by your physician or neurologist with credentials and affiliated institution.

Please visit the CACNA1A Foundation Website for a sample plan
(https://www.cacna1a.org/hm-resources-action-plan)

SPECIAL THANKS TO:

Shannon Babineau, MD Anne Ducros, MD, PhD

- The CACNA1A Foundation is a family-led 501(c)(3) nonprofit organization
- We are a global community dedicated to creating awareness, supporting families and finding a cure for *CACNA1A* genetic variants.
- Our website contains resources for families and professionals who want to learn more about CACNA1A variants
- Please visit our website for more information about how to join us and make a difference. https://www.cacna1a.org/

The information in this brochure is evolving as our medical experts make progress on a consensus for treatment guidelines. Please visit our website for updated guidelines and additional resources, including appropriate treatments and medications. The contents of this guide are not intended to substitute for professional medical advice, diagnosis or treatment. Please consult your physician for personalized medical advice.

CACNA1A FOUNDATION

GUIDE TO CACNA1A-RELATED HEMIPLEGIC MIGRAINE

INFORMATION FOR FAMILIES & MEDICAL PROVIDERS

WWW.CACNA1A.ORG

WHAT IS A HEMIPLEGIC MIGRAINE (HM)?

First, let's explain a **migraine** – A migraine is moderate to severe headache that is focal (in one part of the head), causes limitation in activity and a need to rest, is usually throbbing, worse with movement, and has light/sound sensitivity or nausea/vomiting. It typically lasts 2 to 72 hours. A **migraine with aura** is a headache preceded by temporary neurological symptoms such as a visual disturbance (blind spot/flashing lights), numbness or tingling of an arm/face/-leg, difficulty with speech (knowing what you want to say but not being able to say words correctly/not understanding words).

A **hemiplegic migraine** is a migraine with aura where the aura includes motor symptoms such as weakness of the face, arm, and/or leg. The distinguishing feature of a hemiplegic migraine is one-sided weakness or paralysis. There may also be other aura symptoms present, such as changes in vision, sensation, and/or speech. Typically, the motor symptoms resolve in under 72 hours, but there are reports of them lasting weeks before returning to normal. Hemiplegic migraines are associated with four genes, one of which is *CACNA1A*.

In a HM the dysfunction of the calcium channel leads to cortical spreading depression (an electrical wave that spreads across the surface of the brain that first activates and then briefly inactivates the nerve cells, leading to an inflammatory response), resulting in temporary changes in perception, including aura symptoms such as visual changes, sensory changes, speech changes, and weakness. It also can trigger a headache. It is important to note that there are some common *CACNA1A* variants associated with a more progressive and severe HM, resulting in uncontrolled seizures, and potentially life-threatening brain swelling.

As a result, *CACNA1A*-related Hemiplegic Migraine is a medical emergency that requires immediate treatment to prevent permanent damage, or even death. It should be treated with the same sense of urgency as a seizure.

Two types of HM associated with CACNA1A

1) Familial hemiplegic migraine occurs from a genetic change that can be passed down through families. *CACNA1A* mutations cause FHM type 1 (FHM1)

2) Sporadic hemiplegic migraine is when there is no family history and the patient is determined by genetic testing to have a de novo (present for the first time in the family) *CACNA1A* variant.

CACNA1A SEVERE HEMIPLEGIC MIGRAINE (CSHM)

Hemiplegic Migraine with Associated Seizures and/or Brain Swelling

In some patients with *CACNA1A* mutations, hemiplegic migraine presents as progressive and severe, resulting in uncontrolled seizures, and/or potentially life-threatening brain swelling. **The most distinguishable feature is stroke-like symptoms with one-sided weakness and/or paralysis.**

Patients who are susceptible to CSHM often have co-morbidities like seizures, dystonia, ataxia, nystagmus and other eye movement disorders. It can be difficult to recognize the differences for caregivers and doctors, especially the first time a CSHM occurs. To better recognize a potential CSHM, refer to www.cacna1a.org/hemiplegic-migraine for caregiver-provided videos.

Triggers may include:

- Minor head trauma
- Seizures
- Emotional or physical stress
- Exertion
- Viral infections
- Lack of sleep
- Menstrual period
- Catheter angiography

Note on minor head trauma: Typically, within an hour of the injury a child might develop a headache, vomiting or a fever.

Note on CSHM with seizures: The weakness on one side of the body becomes evident after seizures are controlled. If a patient requires sedation to control seizures, a CSHM may not be detectable and should be treated if at all suspected. For those whose CSHM is triggered by seizures, it can be difficult to differentiate when seizure has stopped and CSHM begins due to some similarities in presentation (such as eye deviation).

Who is at risk for CSHM?

It appears these events are linked with certain Gain of Function variants (or mutations) in the *CACNA1A* gene including most commonly: p.S218L, p.R1349Q, or p.V1396M. Additional variants will be listed on the CACNA1A Foundation website as they are confirmed. These types of attacks often start in early childhood, including as young as the first year of life. When properly managed, damage from CSHM can be prevented.

If a patient has a *CACNA1A* genetic variant and presents with the following systems, immediate treatment is required:

- One-sided weakness and/or paralysis is the most distinguishable symptom
- Eye deviation
- Increased nystagmus (more than the patient's baseline nystagmus)
- Decreased responsiveness and/or altered consciousness
- Vomiting
- Development of a fever
- Seizures

TREATMENTS

Seek emergency treatment at the first sign of symptoms.

Rapid administration of treatment medications below should be administered prior to obtaining imaging. Patients should receive a 3-5 day round of treatment to ensure the CSHM has resolved.

For CSHM: 1: Verapamil 2: Acetazolamide 3: Ketamine

For cerebral edema: 1: Steroids (methylprednisolone) 2: Hypertonic saline 3: Mannitol

For Pain management and nausea: 1: Motrin 2: Ketorolac 3: Zofran

Medications to avoid: 1: Standard migraine medications, which can reduce blood flow (like ergotamine and dihydroergotamine) 2: Triptans

*Newer migraine rescue medications (CGRP antagonists and ditans) do not have as much concern for changing blood flow and may be considered in the future for those with HM, but they are too new to offer any information at this time.

Prophylactic options: 1: Verapamil 2: Flunarizine 3: Acetazolamide (be aware of acidosis risk) 4: Topamax 5: Supplements to consider: Riboflavin, CoQ10 6: If minor head trauma is a known trigger, a protective helmet may be worn