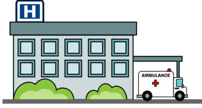

Institution Name
Address
Phone Number
Email

Action & Emergency Plan for *CACNA1A*-related Hemiplegic Migraine

Date:

Information on the Patient
Patient's Name: Patient's DOB: Diagnosis: Allergies: Current Medications/Devices/Diets/Treatments:
About *CACNA1A*-related Hemiplegic Migraines
(Patient's Name) is followed by (name of institution) for (Patient's Name) has had at least one significant hemiplegic migraine episode in the past and the following plan may outline a suggested action plan for the family and the evaluation in a hospital setting. **Hemiplegic migraine episodes in *CACNA1A*-related disorders are NOT typical migraine symptoms and require urgent treatment and evaluation.** These episodes represent a distinct neurological dysfunction that may be progressive and severe, resulting in status epilepticus, and potentially life-threatening brain swelling. -If known, include a description of how the patient's hemiplegic migraines present.
Home Treatment
- Diamox 125 mg - Ibuprofen/Motrin 10 mg/kg - Diastat/rescue benzodiazepine if patient had seizures with HM episode in past
Emergency Room Treatment
If hemiplegic migraine episode persists or patient has not returned to baseline: -ICU admission due to concern of emerging brain swelling - MRI Brain, head CT if MRI brain is not available to assess early brain swelling - Continuous video EEG - Consider I.V. verapamil, 0.1-0.3 mg/kg/dose with maximum 5 mg/dose, Administered with continuous EKG monitoring, have I.V. calcium readily available. - Consider I.V. methylprednisolone 20-30 mg/kg/day (max 1 g) - Consider additional treatment as per migraine pathway: metoclopramide 0.2 mg/kg IV/PO, ketorolac 0.5 mg/kg I.V., Ibuprofen 10 mg/kg PO, methylprednisolone 2 mg/kg I.V. (if lower

SAMPLE

steroid dose is preferred)
- Aggressive seizure treatment according to separate seizure action plan
- Rule out alternative causes of hemiplegia (e.g. vascular stroke), the suggested plan should not prevent work-up of potentially treatable alternative etiologies

If hemiplegic migraine episode has subsided:
- Consider admission for observation. Rationale for this is that prior presentation may have been mild and not predictive for future severity
- Severe presentations with edema have been observed for a specific *CACNA1A* variant including as p.S218L, p.R1349Q, or p.V1396M, which require additional scrutiny

Upon hospital/ICU admission, consider the following:
- Observation for at least 48-72 hours due to concern of delayed brain swelling
- Further treatment upon ongoing disease presentation. Relatively good outcomes with minor deficits have been observed in *CACNA1A*-related disorders even with extreme hemispheric swelling, which should not preclude aggressive treatment with dexamethasone, hypothermia, or decompression craniectomy.

SAMPLE

Physician Signature

Attending Physician's Name
Credentials
Institution Name