



:: Hemiplegic Migraine (HM)



- *This document is a translation of the French recommendations drafted by Dr. Anne Ducros and Dr Gaële Comte, reviewed and published by Orphanet in 2010.*
- *Some of the procedures mentioned, particularly drug treatments, may not be validated in the country where you practice.*

Synonyms:

Migraine with motor aura

Definition:

- ▶ **Hemiplegic Migraine (HM)** is a rare variety of migraine with motor aura (migraine accompanied by transient motor weakness). The family history will allow a distinction to be made between cases of **familial hemiplegic migraine (FHM)**, in which at least one 1st or 2nd degree blood relative suffers from the same crises, and cases of **sporadic hemiplegic migraine (SHM)**, in which no blood relative is affected.
- ▶ **typical HM crises consist of a motor deficit** that is **always associated with at least one other sign of aura**, the most common being sensory, visual and speech problems. Symptoms referred to as "basilar" are also common: vertigo, instability and tinnitus.
- ▶ **Severe HM crises** can develop in both FHM and SHM and consist of **protracted motor deficit, confusion or coma, fever and epileptic crises**.
- ▶ Between consecutive HM crises, 80-90% of patients are asymptomatic. In 10-20% of cases, the clinical picture may include permanent cerebellar signs (nystagmus, ataxia, dysarthria) and, less commonly, epilepsy and intellectual deficiency.

Further information:

[See the Orphanet abstract](#)

Pre-hospital emergency care recommendations

Call for a patient suffering from hemiplegic migraine

Synonyms

- ▶ migraine with motor aura, HM,
- ▶ in the form of:
 - familial hemiplegic migraine, FHM
 - sporadic hemiplegic migraine, SHM

Mechanisms

- ▶ migraine accompanied by an aura featuring motor deficit associated with at least one other symptom (such as visual / sensory problems, aphasia, vertigo, tinnitus)

Specific risks in emergency situations

- ▶ severe crises: protracted motor deficit, hyperthermia (sometimes intense), confusion, coma with inherent complications (agitation, risk of aspiration, respiratory failure), epileptic crises, status epilepticus
- ▶ a minor head injury or a stress situation can trigger a crisis

Commonly used long-term treatments

- ▶ treatment for crises:
 - paracetamol
 - nonsteroidal anti-inflammatory drugs (ketoprofen, aspirin)
 - neuroleptic anti-emetic (metoclopramide)
- ▶ basic treatment (daily):
 - effective anti-epileptic agents for preventing migraine with aura (topiramate, sodium valproate, lamotrigine)
 - other anti-migraine treatments (beta-blockers, anti-calcium agents, tricyclics)

Complications

- ! – rule out hypoglycaemia
- if there is no definite diagnosis, screen for a vascular or infectious cause

Specific pre-hospitalisation medical care

- ▶ there is no effective treatment for aura
- ▶ do not administer triptans
- ▶ do not administer nimodipine (exacerbation of the aura during severe crises)
- ▶ no contra-indication to the use of usual anaesthetic products
- ▶ orientation in line with severity: Accident & Emergency Department with neurological opinion for typical crises; Resuscitation Department for severe crises

Further information

- ▶ The migraine Trust: <http://www.migrainetrust.org>
- ▶ Please visit www.orpha.net and type the name of the disease → in the summary page click on “Expert centres” on the right tab → select “United Kingdom” in the “Country” field in the Expert centres page.

Recommendations for hospital emergency departments

Emergency issues and recommendations

- ▶ [Typical HM crises](#)
- ▶ [Severe HM crises](#)

1. Typical HM crises

▶ Emergency diagnostics

Diagnosis by elimination - only feasible by means of an interview.

- History: the diagnosis can only be made if the patient:
 - is familiar with his/her illness and/or
 - gives a clear report of onset of at least one identical crisis.
 - is capable of speaking (child and/or aphasia: interview the next-of-kin).
 - diagnosis is helped if the patient is a card-carrier ([Annex 1](#)).
- Description of the crisis:
 - Most commonly, the aura starts with deteriorating **visual, then sensory problems** with ascending **unilateral paraesthesia** extending as far up as the face,
 - **Followed by** onset of **speech problems** and **motor deficit** (hemiparesis more commonly than hemiplegia).
 - **neurological deficit** lasts from **10 minutes to several hours**.
 - **headaches** develop **during or after the deficit** and are often accompanied by nausea, vomiting, photophobia and phonophobia.
- Immediate action: screen for and treat hypoglycaemia (differential diagnosis)
- In A+E:
 - Confirm the diagnosis of HM crisis by **re-interviewing** the patient and/or his/her next-of-kin;
 - No additional investigations needed if a typical HM crisis is diagnosed.
 - **Isolate the patient in a calm and dark place.**

▶ Immediate treatment

- Aura:
 - **No aura treatment has proved genuinely effective.** It is better to leave patients to rest in a calm setting until they have recovered from their deficit.
 - A few publications, each dealing with a very small number of cases, point to improvement in the aura following administration of intranasal ketamine or i.v. naloxone.
- **Treat headaches as soon as possible, with the aim of preventing pain;** this also applies during the aura, if the headache has not yet developed.
 - NSAID (ketoprofen 150 mg) p.o., or aspirin 1g
 - **In the event of severe nausea:** ketoprofen 100 mg and metoclopramide 10 mg, administered rectally
 - **If there is already a severe headache with vomiting:** i.v. infusion: Perfalgan® 1g and metoclopramide 10 mg

2. Severe HM crises

▶ Emergency diagnostics

- **Very difficult diagnosis**, even in patients known to suffer from FHM or SHM.
- **Screen for some other vascular or infectious cause** (additional investigations). **This is essential.**
- **Severity criteria and complications:**
 - **Deep coma**
 - **Respiratory failure**
 - **Hyperthermia** (possibly as high as 41°C)
 - **Total neurological deficit:** hemiplegia, mutism, swallowing problems
 - **Major confusion, with agitation and hallucinations**
 - **Vomiting** (risk of aspiration)
 - **Epileptic crises and, sometimes, status epilepticus** - partial or generalised
- Emergency investigations:
 - Clinical investigations: (immediate action, then regular)
 - **Level of consciousness:** Glasgow scale (may drop as low as 3)
 - **Cardiorespiratory monitoring:** respiratory rate, saturation, pulse and BP, pulmonary auscultation (obstruction or aspiration)
 - **Temperature:** hyperthermia, possibly as high as 41°C
 - **Extent of the deficit:** hemiplegia, mutism, swallowing problems
 - paraclinical investigations: these allow differential diagnosis and screening for factors that exacerbate severe HM crises (see [table](#))
 - Immediate action: screen for hypoglycaemia (differential diagnosis)
 - Laboratory blood and urine tests: **blood sugar, electrolytes, FBC, CRP, liver function test, calcaemia, urine test strips, screen for toxic substances in the blood and urine**
 - Brain scan: **plain CT brain scan at the very least**; an MRI brain scan, plain and with contrast, is better
 - **Lumbar puncture in cases of fever:** to be done **after imaging**
 - **Chest X-ray** (where there is fever and/or breathing difficulty)
 - Electrocardiogram (**ECG**)
 - Electroencephalogram (**EEG**) in suspected epileptic crises or if there is confusion, to screen for non-convulsive status epilepticus

▶ Immediate treatment

Treatment is entirely symptomatic.

- **Nasogastric tube**
- **Peripheral venous line**
- Aura: no treatment has provided proof of its efficacy.
- Hyperthermia: i.v. Perfalgan[®] 1g, every 8 hours
- **Headaches:** i.v. Perfalgan[®] with **metoclopramide if there is nausea**, sometimes accompanied by ketoprofen 100 mg every 12 hours.
- **Epileptic crises and status epilepticus:** i.v. and/or oral **anti-epileptic agent**. Proceed with the same treatment as for other causes of epileptic crises

Orientation

- ▶ Where?
 - **Typical crises:** transport to the **nearest Accident & Emergency Department (A+E)**. Keep under observation until recovery from the neurological deficit then return home
 - **Severe crises with no respiratory failure:** transport to the **nearest A+E Department**, admit to a Neurology Department with a Continuous Care Unit
 - **Severe crises with respiratory failure:** transport to **A+E ("crash team")** then transfer to the Medical Resuscitation Department
- ▶ When? As quickly as possible.
- ▶ How? **For severe crises with fever, confusion, coma, respiratory failure:** ECG, oxygen, peripheral venous line

Drug interactions and contraindications

- ▶ **Avoid angiography involving the use of a line to inject iodine:** any angiography (coronary, brain) **can trigger a severe crisis** with coma and must, therefore, be avoided. **In the absence of any alternative procedure**, the patient must be kept under observation **for 24 hours after the procedure to detect symptoms of a severe crisis**
- ▶ **Triptans:** these remain **contraindicated** even though some authors point to their safety and efficacy in tackling headaches during HM crises. **They must never be used during the aura** (typical or protracted).
- ▶ **Nimodipine:** this can **exacerbate a protracted aura** during a severe crisis

Precautions for anaesthesia

- ▶ No contraindication linked to HM.

Preventive measures

- ▶ Avoid the trigger factors for HM crises (typical or severe): "stress" and head injuries, even minor.

Additional therapeutic measures and hospitalisation

- ▶ **Prevention of deep vein thrombosis:** low molecular-weight heparin (**LMWH**) pending restoration of mobility.
- ▶ **Nasogastric feeding** until normal swallowing has been restored
- ▶ **Motor deficit:** daily physiotherapy as soon as possible; continue after discharge
- ▶ **Aphasia:** speech therapy as soon as possible, then at home
- ▶ **Pressure area care:** positioning, mobilisation, care at points of contact

Organ donation

- ! In theory, the disease allows this but it is highly unusual for the condition of the patient to deteriorate to the point of triggering brain death.
- Organ donation is still possible, even if the individual is brain dead (e.g. after a CVA).

Emergency telephone numbers

- ▶ Please visit www.orpha.net and type the name of the disease → in the summary page click on "Expert centres" on the right tab → select "United Kingdom" in the "Country" field in the Expert centres page.

Documentary resources

- ▶ Ducros A. Familial and sporadic hemiplegic migraine. Rev Neurol (Paris). 2008 Mar;164(3):216-24. Epub 2008 Mar 11.

Annexes

1- Specimen card to be issued to patients

Patient Card - Hemiplegic Migraine	
Name:	_____
Forename:	_____
Date of birth:	_____
Person to be notified (relative, friend):	_____ Mobile: _____
Usual treatment:	_____
Neurologist:	_____ Telephone: _____
<p>I suffer from hemiplegic migraine. During a crisis, I experience motor problems (paralysis of one or more limbs), sensory problems (numbness, loss of feeling in one or more limbs), and speech and vision problems. These neurological problems can last for a few minutes up to several hours. They are followed by headaches with nausea and vomiting.</p> <p>If I am incapable of moving or speaking or if I have lost consciousness: ring 999 so that I can be taken to the nearest A+E Department.</p>	

2- Table

Results from additional investigations during a severe FHM/SHM crisis	
CT / MRI scan	<ul style="list-style-type: none">- During the crisis: normal brain or cerebral oedema with swelling of the cortical ribbon- Check-up after a few weeks: normal cerebral parenchyma (no sequelae)- Sometimes (<15%) cerebellar atrophy, predominantly affecting the anterior part of the vermis, leaving the brainstem intact (permanent abnormality, during crises and between them)
EEG	<ul style="list-style-type: none">- Slow diffuse waves predominantly in the contralateral hemisphere to the deficit; sometimes spiked periodic waves or dysrhythmia.- Abnormalities that can last for weeks after a severe crisis.
CSF	<ul style="list-style-type: none">- Normal or aseptic lymphocytic meningitis ($12-290/\text{mm}^3$), sometimes of mixed formulation or predominantly polynuclear.- Slightly raised CSF protein content (up to 1 g /L)- Normal CSF glucose level- Sterile fluid (negative cultures)
Transcranial Doppler	<ul style="list-style-type: none">- Widespread or localised acceleration of intracranial speeds

These recommendations have been compiled in collaboration with Dr Anne Ducros at the Emergency Treatment Centre for Headaches and at the Reference Centre for Rare Vascular Disorders of the Central Nervous System and of the Retina (CERVCO), Lariboisière Hospital, Paris, and with Dr Gaële Comte, SAMU-69, Lyons

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