

# Medical card / Emergency

**The owner of this card has:**

## **Moyamoya cerebral angiopathy**

*It is recommended that the owner holds this card permanently.*

Moyamoya angiopathy is a rare chronic intracranial arteriopathy occurring in children and adults leading to cerebral infarction and cerebral hemorrhage. It associates progressive steno-occlusive lesions of the arteries of the Circle of Willis with a compensatory network of fragile neovessels. Cerebral ischemia is often related to hemodynamic changes; precipitating factors such as hyperventilation or a drop in blood pressure are sometimes identified. Moyamoya angiopathy may be isolated (moyamoya disease) or may be associated with a local or general condition which is acquired or genetic in nature (Moyamoya syndrome). The hereby proposed recommendations apply equally to both entities.

**More information about diagnostic and care protocol in French:**

[https://www.has-sante.fr/portail/upload/docs/application/pdf/2016-08/pnds\\_-\\_maladie\\_et\\_syndrome\\_de\\_moyamoya\\_de\\_lenfant\\_et\\_de\\_ladulte\\_2016-08-11\\_11-35-20\\_513.pdf](https://www.has-sante.fr/portail/upload/docs/application/pdf/2016-08/pnds_-_maladie_et_syndrome_de_moyamoya_de_lenfant_et_de_ladulte_2016-08-11_11-35-20_513.pdf)

*This card should be filled and updated by the patient's physician, in the presence and with the agreement of its owner. This document is subject to medical confidentiality and no one may require its disclosure without the authorization of the owner or his/her legal representative.*

Edited by



**ASSOCIATION** 2021  
**TANGUY MOYA-MOYA**

# Handling emergency situations (1)

- ▶ **A cerebrovascular accident (transient ischemic attack, ischemic stroke, or hemorrhagic stroke) is the most common complication in patients with moyamoya. In this situation, treatment priorities are:**
  - Close monitoring of blood pressure, heart rate, oxygen saturation, and temperature. Blood pressure monitoring is particularly important as sudden or significant decreases in blood pressure can rapidly worsen neurological status.
  - In the case of sudden focal neurologic deficit, cerebral imaging is key in diagnosing ischemic or hemorrhagic stroke. Cerebral infarction in the acute phase is best detected by MR imaging.
  - If MRI is unavailable or in the case of altered consciousness, urgent brain CT scan can diagnose cerebral hemorrhage but has limited sensitivity in confirming ischemic stroke.
- ▶ **In case of cerebral ischemia (transient ischemic attack or ischemic stroke), it is recommended to:**
  - Accept elevated blood pressure, unless it is clinically poorly tolerated;
  - Avoid sudden lowering of the patient's blood pressure;
  - Use normal saline infusion if volume expansion is required (hypotonic fluids are not appropriate);
  - Maintain the patient in supine position without head elevation;
  - Correct fluid and electrolyte imbalances, hyperthermia, pain (especially in infants to avoid hyperventilation from crying), hypocarbia, hypovolemia;

## Handling emergency situations (2)

- Start antithrombotic treatment with antiplatelet therapy (for example IV aspirin in adults). Adults and post pubertal adolescents who are immobilized or have a lower limb motor deficit require deep vein thrombosis prevention with low-molecular-weight heparin;
- Avoid:
  - anticoagulation at therapeutic dose which increases the risk of cerebral hemorrhage;
  - thrombolytic therapy which is not indicated in adults with cerebral infarction related to moyamoya angiopathy.
- ▶ **When cerebral or subarachnoid hemorrhage is diagnosed, it is recommended to:**
  - Seek urgent neurosurgical review;
  - Tolerate elevated blood pressure except if  $\geq 180 / 105$  mm Hg in adults or +2SD for age in children, and in all cases avoid sudden decreases in blood pressure;
  - Correct fluid and electrolyte imbalance, hyperthermia, pain, hypocarbia, hypovolemia ;
  - Avoid any antithrombotic therapy other than deep vein thrombosis prevention in adults and post pubertal adolescents who are immobilized or have a lower limb motor deficit. In this case, low molecular weight heparin can be started on day 2 if initial hemorrhage remains stable.

*More information can be found on Orphanet Urgences website:  
[http://www.orpha.net/consor/cgi-bin/Disease\\_Emergency.php?lng=FR](http://www.orpha.net/consor/cgi-bin/Disease_Emergency.php?lng=FR)*

# Medical Precautions

## Therapeutic recommendations

- ▶ **Use antihypertensive drugs with caution** (starting or modifying treatment should be done with caution, and in collaboration with the patient's treating medical team). Beware of the risk of aggravating cerebral hypoperfusion potentially causing further ischemic complications. Do not use antihypertensive drugs in an emergency unless systolic BP is  $\geq 230/130$  mmHg in adults or is poorly tolerated (treat if evidence of cardiac decompensation).
- ▶ **Anticoagulants are not recommended** for the prevention of cerebral ischemia (demonstrated a **lack of clinical efficacy** and an increased **risk of cerebral hemorrhage**). Anticoagulants should be used only when strongly indicated (confirmed deep vein thrombosis, pulmonary embolism...), and preferably for a limited period after the risk of hemorrhage has been estimated with the referring physician.
- ▶ **Avoid using vasoconstrictive drugs** because of the risk of worsening cerebral hypoperfusion. Special attention should be paid to ENT decongestants (including medications given by nasal route) and antimigraine drugs (triptans and ergot derivatives). See drug list in the full moyamoya protocol (weblink on page 1 of this card).
- ▶ **Acute dehydration (e.g. in case of gastroenteritis)**, especially in infants, should be prevented by rapid implementation of **appropriate rehydration solutions**, possibly including intravenous fluids.
- ▶ **Avoid hyperventilation** situations (hyperpnoea test during EEG recording is contra-indicated) as hypocarbia leads to vasoconstriction.

## Anesthetic precautions

- ▶ **The administration of general or local anesthesia, even for minor surgery, should be prepared and thoroughly monitored**, in collaboration with experienced staff who are familiar with moyamoya angiopathy and its potential complications.
- ▶ Both anesthesia and the surgical procedure pose a **major risk of neurologic injury** due to the hemodynamic changes they may induce.
- ▶ **Any drop in blood pressure**, even brief in duration, **should be avoided**.
- ▶ All measures should be taken to prevent and aggressively treat pain, hypo or hypertension, hypovolemia, hypoxia, hypocarbia, or any metabolic derangements in the perioperative period.
- ▶ Regional anesthesia should be the technique of choice in moyamoya patients.

## Sports and physical activity safety precautions

- ▶ **Patients with moyamoya should avoid:**

Any activity with **sudden change in body temperature** (sauna, Scottish bath) or **hyperventilation**.

Contact and extreme sports with either a **risk of head trauma** (fighting sports, rugby ...) or where **fainting would be hazardous** (scuba diving, unsupervised swimming, parachuting, uninsured climbing ...).

Apart from these situations, physical activity, including competitive sports or playing musical wind instruments, can be practiced if well tolerated. In children, leisure sports are encouraged over competitive sports. A school certificate outlining these recommendations can be sought.

# Patient's information

**First name:** .....

**Last name:** .....

**Birth date:** .....

## 1/ Clinical history:

- Transient ischemic attack or ischemic stroke
- Cerebral or subarachnoid hemorrhage
- Epileptic seizures
- Cognitive disorder
- Abnormal movements

**Others:** .....

**Surgical revascularization:**      No                       Yes

**Surgeon's name and hospital:** .....

.....

## 2/ Associated conditions (including allergies, contraindications):

.....

## 3/ Current treatment:

- Antithrombotic treatment                     

- Antiepileptic treatment                     

- Other: .....



# Referral centers – More information

## Reference center for adult patients



Tel : +33 1 49 95 25 91

[www.cervco.fr](http://www.cervco.fr)

## French center for pediatric stroke



Email : [AVCenfant@chu-st-etienne.fr](mailto:AVCenfant@chu-st-etienne.fr)

[www.cnravenfant.fr](http://www.cnravenfant.fr)

## Parents' association



Email : [Atmmcontact@yahoo.fr](mailto:Atmmcontact@yahoo.fr)

[www.tanguy-moya-moya.org](http://www.tanguy-moya-moya.org)

## For more information

orphanet

The portal for rare diseases

[www.orphanet.org](http://www.orphanet.org)

Maladies Rares  
Info Services

0 810 63 19 20

N° Azur, prix appel local

*This medical card*

*was edited by the Tanguy Moya-Moya Association.*

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