




**European
Reference
Network**

for rare or low prevalence
complex diseases

 **Network**
Vascular Diseases
(VASCERN)

VASCERN DO'S AND DON'TS FACTSHEETS
FOR RARE VASCULAR DISEASE PATIENTS
FACING FREQUENT SITUATIONS

Moyamoya Angiopathy



VASCERN

VASCERN, the European Reference Network on Rare Multisystemic Vascular Diseases, is dedicated to gathering the best expertise in Europe in order to provide accessible crossborder healthcare to patients with rare vascular diseases (an estimated 1.3 million concerned). These include arterial diseases (affecting aorta to small arteries), arterio-venous anomalies, venous malformations, and lymphatic diseases.

VASCERN currently gathers 48 expert teams from 39 highly specialized multidisciplinary HCPs, plus 6 additional Affiliated Partner centers, coming from 19 EU Member States, as well as various European Patient Organisations, and is coordinated in Paris, France.

Through our 6 Rare Disease Working Groups (RDWGs) as well as several thematic WGs and the ePAG (European Patient Advocacy Group), we aim to improve care, promote best practices and guidelines, reinforce research, empower patients, provide training for healthcare professionals and realise the full potential of European cooperation for specialised healthcare by exploiting the latest innovations in medical science and health technologies.

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Moyamoya Angiopathy

Abbreviations

MMA: Moyamoya Angiopathy

MMD: Moyamoya Disease

TIA: Transient Ischemic Attack

NEUROVASC-WG: Neurovascular Diseases Working Group

Introduction

These factsheets are based on existing French factsheets which have been reviewed and adjusted by the experts of the VASCERN NEUROVASC-WG.

The NEUROVASC-WG agrees with the recommendations but wishes to emphasize that these are recommendations made by consensus at expert level. We would recommend that these factsheets be used as a guide to implement locally agreed policies.

These factsheets are meant for patients as well as for caregivers. Implementing these recommendations should go hand in hand with strategies to educate patients about medical situations where specific care is required and about relevant symptoms and how to act when they occur.

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Anaesthesia

The haemodynamic consequences of surgery and general anaesthesia can play an important role in the occurrence of perioperative cerebrovascular complications in patients with Moyamoya angiopathy (MMA).



WHAT IS RECOMMENDED

- Careful monitoring and control of clinical or metabolic disturbances (pain, arterial hypotension including of orthostatic origin, hypovolemia, hypocapnia, and hypertension) are recommended during anaesthesia.
- Contact the patient's reference centre before a general anaesthesia, independently of the nature of the surgical treatment.
- The anaesthesia procedure with the lowest risk of causing blood pressure fluctuations should be preferred.



WHAT YOU SHOULD NOT DO

- Expose the patient to sudden and/or wide blood pressure fluctuations.

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Antihypertensive drugs

Antihypertensive drugs should be used with caution in patients with MMA, due to the risk of worsening cerebral hemodynamic impairment. Significantly elevated blood pressure can increase the risk of cerebral haemorrhage.



WHAT IS RECOMMENDED

- In all patients, a 24 hours ambulatory blood pressure monitoring should be carried out. In case of hypertension, blood pressure should be lowered always carefully and gradually.
- The target values should be determined on the basis of the individual prevalent values and should not be based on normal values of healthy controls.



WHAT YOU SHOULD NOT DO

- Prescribe antihypertensive drugs in case of TIA or at the acute stage of ischemic stroke without knowing haemodynamic status.



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Antiplatelet Agents

In patients without prior cerebral haemorrhage, sustained antiplatelet therapy is suggested for prevention of embolic stroke without increasing the risk of cerebral haemorrhage.



WHAT IS RECOMMENDED

- Prescribe antiplatelet drugs (aspirin in first intention) only in case of ischemic cerebrovascular event or silent ischemic lesion on MRI.

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Anticoagulants

The use of anticoagulants is not recommended in patients with MMA due to the increased risk of cerebral haemorrhage. Their long-term efficacy on the occurrence of ischemic stroke has not been demonstrated.



WHAT IS RECOMMENDED

- Prescribe anticoagulants only in case of indication without alternative (i.e. thromboembolic complications).
- In case of atrial fibrillation, left atrial appendage closure should be considered.



WHAT YOU SHOULD NOT DO

- Prescribe anticoagulants to prevent ischemic stroke related to MMA.



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Thrombolysis

There is no sufficient data to make any recommendation about thrombolysis in case of ischemic stroke related to MMA.



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Antiepileptic Drugs

The use of antiepileptic drugs is possible for patients with MMA.



WHAT IS RECOMMENDED

- Prescribe antiepileptic drugs for epileptic seizures.

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Contraception

The use of contraceptive pills is a prothrombotic risk factor.



WHAT IS RECOMMENDED

- It is preferable to use progestogen-only pill (without oestrogens) rather than the classical combined oral contraceptive pill.



WHAT YOU SHOULD NOT DO

- The combination of oestrogen pills with smoking should be particularly avoided.



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Menopause

The treatment of menopause-related symptoms with hormones can increase the thrombotic risk, in addition to the risk related to MMA.



WHAT IS RECOMMENDED

- Prescribe hormonal treatment only if they lead to a real benefit on menopause-related symptoms and whenever possible for a limited period.

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Pregnancy and Delivery

Pregnancy is possible in patients with MMD, as long as it is followed carefully by a medical team which is alert to the complications of MMA.



WHAT IS RECOMMENDED

- Discuss any planned pregnancy with the doctor prior to conception.
- Contact the patient's reference centre before delivery, to ensure a correct follow-up.
- Careful monitoring and control of clinical or metabolic disturbances (pain, hypotension, hypertension, hypovolemia, and hypocapnia) are recommended during delivery.
- The anaesthesia and delivery procedures with the lowest risk of causing blood pressure fluctuations should be preferred. Normal delivery or caesarean section should be discussed depending on the haemodynamic status on an individual basis.

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Vasoconstrictors

The use of vasoconstrictors (such as nasal vasoconstrictors, triptans and ergot derivatives) should be avoided due to the risk of worsening cerebral hemodynamic impairment.



WHAT IS RECOMMENDED

- Non vasoconstrictive drugs should be preferred for migraine.



WHAT YOU SHOULD NOT DO

- Prescribe vasoconstrictors in patients with Moyamoya.

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Exercise

A regular practice of a sport and/or a physical activity is recommended in patients with MMA.



WHAT IS RECOMMENDED

- Regular sports and/or physical activity can be considered if they are well tolerated.
- In children, it is usual to favour leisure sports over competition.



WHAT YOU SHOULD NOT DO

- Activities with sudden changes in body temperature (sauna, Scottish baths) or hyperventilation.
- Violent and extreme sports with risk of head trauma (combat sports, rugby,...) especially after bypass surgery.
- Sport activity during which the occurrence of a loss of consciousness can be dangerous (scuba diving, unsupervised swimming, parachuting, uninsured climbing).



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Organ & Tissue Donation

Organ donation is generally possible in patients with Moyamoya disease, depending on their particular medical state. In the case of patients with Moyamoya syndrome, the damage of the organs concerned by the associated condition should be evaluated.



WHAT IS RECOMMENDED

- Donation of blood vessels should be considered with caution.

Editorial Board/ Contributors

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