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complex diseases

 **Network**
Vascular Diseases
(VASCERN)

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

Cerebral Autosomal Dominant Arteriopathy with Sub-cortical Infarcts and Leukoencephalopathy (CADASIL)



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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Cerebral Autosomal Dominant Arteriopathy with Sub-cortical Infarcts and Leukoencephalopathy (CADASIL)

Abbreviations

CT: Computed tomography

MRI: Magnetic resonance imaging

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.

Thrombolysis

Intravenous thrombolysis is not recommended in CADASIL patients with lacunar stroke.



WHAT IS RECOMMENDED

- Intravenous thrombolysis should be avoided in CADASIL patients with lacunar stroke (ischaemic) related to the disease, due to the potential increased risk of intracerebral haemorrhage.

Antiplatelet Agents

Antiplatelet agents are often used as a preventive treatment in patients with CADASIL after an ischemic event or in the presence of small deep infarcts on imaging. However, the efficacy of antiplatelets in stroke prevention in CADASIL is not proven.



WHAT IS RECOMMENDED

- Use antiplatelet agents (aspirin or clopidogrel) only after the occurrence of clinical ischemic events (transient ischemic attack or completed stroke). Discuss this indication in presence of covert cerebral infarcts (symptomatic or not) on CT-scan or MRI.



WHAT YOU SHOULD NOT DO

- Prescribe antiplatelet drugs in the absence of cerebral infarctions on MRI or clinical ischemic events.

Anticoagulants

The use of anticoagulants is generally not recommended for secondary stroke prevention in patients with CADASIL, when the stroke event is presumably related to the genetic disease, since the disease can promote the occurrence of haemorrhagic stroke.

However, the use of anticoagulant is not contraindicated in case of another strong indication (e.g. atrial fibrillation, pulmonary embolism).



WHAT IS RECOMMENDED

- Prescribe anticoagulants only under formal indications, i.e. thromboembolic complications or atrial fibrillation, and re-evaluate regularly the risk-benefit ratio.



WHAT YOU SHOULD NOT DO

- Prescribe anticoagulants to prevent cerebral ischemic manifestations (clinical and/or neuroimaging) related to the disease.

Statin Therapy

Statin therapy is not recommended in CADASIL patients to prevent stroke related to disease.



WHAT IS RECOMMENDED

- Prescribe statins only in presence of a clear indication distinct from the occurrence of ischemic stroke manifestations presumably related to the disease.



WHAT YOU SHOULD NOT DO

- Prescribe statins in CADASIL patients after an ischemic stroke presumably related to the disease, in the absence of another indication.

Anaesthesia

Anaesthesia, both local and general, should be used with caution in patients with CADASIL. Exposure to large variations of blood pressure during anaesthesia may increase the risk of ischaemic stroke and clinical worsening.



WHAT IS RECOMMENDED

- Haemodynamic stability and avoidance of large drop(s) of blood pressure.



WHAT YOU SHOULD NOT DO

- Use anaesthetics without monitoring and correcting blood pressure variations.

Digital Subtraction Angiography

The use of digital subtraction angiography is not recommended in CADASIL since severe complications such as migraine with prolonged aura or a diffuse and severe encephalopathy are possible.



WHAT IS RECOMMENDED

- CT angiography or ultrasound examination for large artery evaluation in CADASIL patients.



WHAT YOU SHOULD NOT DO

- Digital subtraction angiography should not be considered as the first line examination of large intracranial arteries for CADASIL. However, it can be considered in some specific situations.

Pregnancy

Pregnancy does not seem to increase the risk of stroke nor trigger the occurrence of any specific manifestation of the disease in women with CADASIL. However, some patients can present with severe attacks of migraine with aura during pregnancy or in the weeks after delivery.



WHAT IS RECOMMENDED

- Regular neurological follow-up of female CADASIL patients during pregnancy.



WHAT YOU SHOULD NOT DO

- Prevention of cerebral ischemic events with heparin or aspirin during pregnancy is not recommended in CADASIL.

Treatment of Migraine

There is no evidence contraindicating the use of triptans in patients with CADASIL suffering from migraine attacks.



WHAT IS RECOMMENDED

- Use triptans or similar vasoconstrictive drugs only as second or third choice of acute treatment of migraine attacks in CADASIL.



WHAT YOU SHOULD NOT DO

- Prescribe drugs with potential vasoconstrictive effects in patients with CADASIL as first line treatment of migraine attacks.

Treatment of Psychiatric Features

Patients with CADASIL can be treated with psycho-effective drugs according to the usual guidelines.



WHAT IS RECOMMENDED

- All treatments can be used if well tolerated and not inducing severe hypotension.



WHAT YOU SHOULD NOT DO

- Use psycho-effective drugs without any follow-up and blood pressure monitoring.

Menopause

There is no data concerning the potential risk of treatments used for menopause-related manifestations.



WHAT IS RECOMMENDED

- The use of hormonal treatments should only be prescribed in the presence of menopause symptoms altering the quality of life.



WHAT YOU SHOULD NOT DO

- Prescribe hormonal treatment in the absence of a menopause symptoms and over an unlimited period.

Organ Donation

Organ donation is possible in patients with CADASIL, since there is no clinically significant extracerebral involvement.

There is no specific recommendation on this aspect.

Editorial Board/ Contributors

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