

# Carotid-cavernous fistulas

Carotid-cavernous fistula is a possible complication of vascular Ehlers-Danlos syndrome. It affects around 10-15% of patients with the syndrome, and consists of a rupture of the intracranial carotid artery into the main collection site of venous blood in the brain. Its spontaneous character is pathognomonic for vascular EDS.



## WHAT IS RECOMMENDED

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- The appearance of a pulsating murmur in the auscultation of the skull or the eye will suggest the presence of a carotid-cavernous fistula. This suspicion is increased when the eye on the same side as the murmur becomes red and/or painful. Swelling around the eye may also be present.
- If a carotid-cavernous fistula is suspected, an angiography /cerebral MRI scan is recommended.
- Management and care in a neurovascular unit is required. Therapeutic occlusion of the fistula is often necessary, in spite of the inherent risk of the procedure. This procedure must be carried out in a specialist centre with the greatest possible level of experience.
- Blood pressure in particular should be closely monitored. Maintain a blood pressure target <130/80 mmHg. A protocol of permissive hypotension is recommended without compromise to organ function.
- Caution with use of inotropes is recommended.
- Caution with use of indwelling catheters is advised.



## WHAT YOU SHOULD NOT DO

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- Delay imaging procedures which alone determine the diagnosis.
- Delay embolization when it is absolutely necessary, as it is a source of significant functional sequelae.