Vascular Ehlers-Danlos syndrome

Peripheral arterial dissection

Peripheral arterial dissections, especially those of the medium-sized arteries, are the most frequent complication of vascular Ehlers-Danlos syndrome. They are spontaneous and must be suspected in the presence of an unexplained pain syndrome.



WHAT IS RECOMMENDED

- Call the national centre of reference or the regional centre of competence to seek advice due to the seriousness of the risk of dissection.
- Quickly implement all necessary actions in order to confirm the diagnosis of vascular Ehlers-Danlos Syndrome.
- Preference should be given to non-invasive examinations (such as MRA or CTA) and medical/conservative treatment whenever possible.
- When an arteriography is necessary (rupture, perforation), particular precaution should be taken when the guide is being moved upwards (due to risk of dissection/arterial perforation) and enhanced monitoring of the femoral insertion point is necessary.
- Arterial ruptures should be treated in preference by embolization.
- 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

- The insertion of stents (except in the case of a life-threatening emergency) should be avoided.
- Consider surgical treatment, other than as a last resort.
- Systematically initiate treatment with anticoagulation and/or antiplatelet agents.