



# Type B intramural hematoma and descending penetrating aortic ulcer

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## Introduction

There is a variety of partly overlapping or even merging acute aortic diseases summarized as acute aortic syndrome, that is, aortic dissection (AD), intramural hematoma (IMH) and penetrating aortic ulcer (PAU). IMH is a disease of the media and defined as aortic wall thickening >5 mm caused by hemorrhage into the media layer without a detectable flow within the media, and without noticeable communications between the layers in the majority of cases (1). Nevertheless, one should be aware that a primary entry tear exists (even though it is not detectable on CTA) and a ruptured atherosclerotic plaque indicates the initial site of IMH (2). Furthermore, PAU is characterized by an atherosclerotic plaque rupture resulting in exposure of the media to varying extent and depth (3). While there is an increasing understanding of the most frequent aortic pathologies such as AD and aortic aneurysms, evidence for the natural course as well as the appropriate treatment of IMH or PAU is rare.

## IMH

The incidence of IMH is reported to be 5–20% of patients with acute aortic syndromes (4). While an IMH is limited to the descending aorta in the majority of the cases and is less frequently associated with malperfusion syndromes, it is more difficult to define the extent of the disease process, especially in the proximal descending aorta or within the aortic arch, making optimal treatment decisions more difficult. Medical treatment, based on accurate blood

pressure control, pain management and imaging follow-up, is standard therapy in uncomplicated scenarios because of the higher rate of disease regression compared to AD. Several clinical (e.g., age and persistent pain), or imaging findings (initial aortic diameter >45 mm, the increase of mean aortic diameter >5 mm/year, wall thickness >10 mm, pleural effusion, aortic ulcer or ulcer-like projection) may necessitate invasive treatment or at least should be considered in the treatment decision-making process (1). When optimal medical treatment is insufficient and endovascular or surgical treatment is required, TEVAR is considered to be standard therapy for the proximal descending aorta (4). However, one should be aware that the proximal landing zone should not be affected by disease and the wall diameter should be regular due to the relevant risk of inducing an iatrogenic AD. Additionally, oversizing should be avoided or at least limited to <10% of the aortic diameter. In cases without a suitable proximal landing zone or in scenarios of retrograde type A progression of the IMH (which would require some type of supra-aortic transposition), the frozen elephant trunk (FET) technique is considered as a safe open surgical treatment option with excellent clinical outcomes as well as remodeling of the descending aorta (5). Moreover, the higher risk of IA endoleaks in IMH patients can be avoided easily by using the FET technique.

## PAU

PAUs of the downstream aorta are often an incidental finding in imaging performed for non-aortic related reasons

and can occur independently or in association with IMH. In these cases, PAU has to be carefully differentiated from ulcer-like projections, which occur after the initial finding of IMH. Unfortunately, little is known about the incidence, natural course, and risk of mortality in patients with PAU. Thus, the risk of mortality, progression to classic AD and risk of aortic rupture is controversial, but certainly relevant (6). There are several findings predicting a complicated course (persistent pain, IMH, periaortic bleeding or a large ulcer size) (1), although an objective cut-off regarding the best choice of treatment is missing. Despite best medical treatment, entire coverage of a localized lesion is indicated where complicating factors are present. Although TEVAR seems to be the treatment of choice, decision making is more demanding than it initially appears. One should be aware that PAU is part of systemic atherosclerotic disease, which often affects access vessels, the abdominal aorta or the coronary arteries. This fact should be carefully considered in order to avoid access vessel complications or treatment related distal embolisation due to an atherosclerotic plaque rupture. Tailored considerations are necessary in each individual patient based on accurate downstream and abdominal aorta computed tomography angiography (including the access vessel), as well as coronary imaging. Consequently, if there are any contraindications or doubts concerning endovascular treatment or coverage of PAU in the distal aortic arch or proximal descending aorta, one should consider total aortic arch replacement using the FET technique. FET has become a well-established option in recent years, especially when additional coronary artery bypass grafting is required (5). In the more distal downstream aorta, open surgical thoracoabdominal replacement can be performed with favorable results in high-volume aortic centers (7).

## Conclusions

Despite accurate imaging and the best medical treatment of acute aortic syndromes affecting the descending aorta, TEVAR is the treatment of choice in eligible patients. Nevertheless, individual clinical and imaging findings may complicate endovascular treatment and require an open surgical approach. In these cases, total aortic arch replacement using the FET technique is the most suitable option for pathologies involving the distal aortic arch or the proximal descending aorta, especially in scenarios with unfavorable anatomy or if any concomitant cardiac procedure is required.

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## Footnote

*Conflicts of Interest:* The authors have no conflicts of interest to declare.

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