The term acute aortic syndrome (AAS) embraces a heterogeneous group of patients with a similar clinical profile of acute presentation and characteristic “aortic pain”. Acute aortic syndrome encompasses classic aortic dissection, less common variants such as intramural hematoma, penetrating aortic ulcer, and iatrogenic or traumatic transection of the aorta. In some patients, AAS may also be caused by symptomatic degenerative aortic aneurysm. Eventually, any one of AAS may progress to frank aortic rupture that will be contained or not. These acute aortic pathologies appear separately, may precede one another and/or just coexist. AAS is the most frequently fatal condition in the spectrum of patients with chest pain. These patients are characterised by “aortic pain” and a long-lasting history of severe hypertension.

Acute aortic dissection is the most common cause of AAS, while the most common predisposing factor for aortic dissection is systemic hypertension. Classic aortic dissection exhibits the presence of an intimo-medial flap and an entrance tear, which typically occurs at sites of highest intra-luminal pressure and wall tension. Type A dissections involve the ascending aorta, the aortic arch and different lengths of the descending and abdominal aorta, while type B dissections involve the descending thoracic aorta distal to the left subclavian artery. Type A dissections are managed surgically, uncomplicated type B dissections are managed medically in most instances, while acute complicated aortic dissections can be treated by stent-grafting.

Intramural hematoma (IMH) lacks a detectable intimal tear/disruption, and has therefore no communication with the aortic lumen, but is confined within the aortic wall. Management of IMH is similar to dissection, although guidelines are not as well established. Penetrating aortic ulcer (PAU) is defined by an ulceration of an aortic atherosclerotic plaque penetrating through the internal elastic lamina into the aortic media and occurs most often in patients with extensive atherosclerotic disease. Penetrating ulcers of the ascending aorta (type A PAU) are rare, but dreadful, requiring urgent surgical repair in most patients. Patients with symptomatic PAUs of the descending aorta, who are not good surgical candidates, are excellent candidates for urgent stent-graft placement.

In the current era, since traditional open surgical procedures share high mortality and morbidity rates, the use of endovascular techniques seem to offer significant survival benefits. Finally, in certain cases, such as uncomplicated type B dissection, or during the pre-operative phase, patients should be managed medically targeting at both clinical and hemodynamic stabilization.