Aortic intramural hematoma (IMH) has been considered a variant or precursor of aortic dissection with no entry tear or false lumen flow; however, the pathophysiological mechanism, risk factors, and evolution are rather different from those of classic dissection. Hematoma forms within the aortic wall as a result of either hemorrhage of the vasa vasorum or, less commonly, an intimal fracture of an atherosclerotic plaque. The marked relationship between IMH and atherosclerotic disease explains the older age of these patients compared with those with aortic dissection, the higher incidence of arterial hypertension, and the tendency for descending aorta involvement (50% to 60%).

Evolution of the IMH in the acute phase may be highly dynamic, with bleeding of the aorta wall increasing progressively, stabilizing or provoking disruption of the intima, which may lead to a classic or localized dissection. This intimal disruption may be seen in the early hours of presentation or some months after the intramural bleeding has occurred. 

First, IMH might regress spontaneously with time. Second, fewer severe cardiovascular complications, valvular aortic regurgitation, and visceral or peripheral ischemia are present. Therefore, IMH almost resembles an aortic dissection, but with a distinct, unique pathological nature.

In patients with uncomplicated type B IMH, the consensus is that medical treatment is recommended. However, the initial treatment strategy for type A IMH remains controversial. Owing to a lower prevalence of the disease, publication updates have been limited to a small number of patients in single hospitals, multicenter registries, or meta-analyses, all of which have limitations. At several centers, particularly in Japan and Korea, patients were observed on medical therapy alone, with timed surgical intervention in cases with complications and favorable outcome, with mortality <10% being reported. However, most Western aortic centers noted that patients who underwent surgery had a significantly better prognosis than those who received medical treatment. Two meta-analyses from Asian and Western series, with an inclusion of >300 type A IMH cases, showed that patients sent for early surgery had slightly lower in-hospital mortality (10.1% to 10.4%) than those treated with early medical treatment (14.4% to 18.4%). Thus, overall agreement on the best strategy for patient management has not yet been reached.

In this issue, Song et al published the largest series of type A IMH reported, 101 cases, collected from a single center. The treatment policy of that institution was initial medical treatment with timed surgical intervention in cases with complications according to clinical assessment and on follow-up imaging studies. Urgent surgery was recommended for hemodynamically unstable patients. Remarkably, urgent operations were performed only in 16 patients with unstable IMH (15.8%). The indication for urgent surgical treatment was not defined clearly because 16 operated cases underwent surgery because of tamponade in 8, syncope in 4, and rapid progression to dissection in 4. Nevertheless, the reason why the remaining 13 patients with tamponade and the 15 patients with syncope were not operated on was not specified. With the use of this strategy, overall hospital mortality was lower in IMH patients than in aortic dissection patients (7.9% versus 17.2%; P<0.03) and was comparable to that of surgically treated aortic dissection patients. This study confirmed some preliminary results that showed that early medical treatment, with closely monitored follow-up, was a reasonable option in the treatment strategy of “stable” type A IMH. However, some points should be considered. The reason why an Asian cohort of patients may fare better with medical management remains unclear. It should be noted that IMH is diagnosed much more frequently among acute aortic syndromes in Japan/Korea (32%) than in European Union/Europe (11%), which has raised the possibility of a genetic or environmental factor. However, other reasons could account for these differences, which might explain the different prognoses of this disease. The significant advances in imaging techniques may have enabled a more correct diagnosis of this condition. Nevertheless, the diagnosis of IMH implies a higher level of expertise than that of aortic dissection. A notable fact is that, unlike aortic dissection, IMH may go unnoticed in the first imaging study, as occurred in 13% of the IMH cases included in the International Registry of Acute Aortic Dissections (IRAD) series. The explanation for this enigma is that, although abrupt disruption of the intima is produced in dissection, IMH is produced by bleeding of the vasa vasorum, which is frequently progressive. Thus, in early

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studies, IMH thickness may be small, and more subtle findings of wall thickening may be overlooked, which are then clearly demonstrated by a second imaging technique. One example of this is that only 5% of acute aortic syndromes were diagnosed as IMH at IRAD referral centers. Thus, it is plausible to think that some benign IMH were not diagnosed in community hospitals and the most “malignant” cases of IMH were referred to specialized centers. In support of this concept, it is noteworthy that in some Western series from single centers or national registries, the incidence of IMH was higher than in the IRAD series, ≈15% of acute aortic syndrome. It seems logical that the inclusion of more subtle cases of IMH, potentially indicative of less severe or more stable cases of IMH, may have led to improved outcomes with medical therapy in Asian series. On the other hand, owing to the early dynamic evolution of IMH, 15% to 20% of cases may lead rapidly to aortic dissection; thus, some of these cases may have been included as aortic

Figure 1. Acute type A IMH (white arrows) with ascending aorta diameter of 45 mm and intramural thickness of 8 mm (left). After 6 days, the ascending aorta dissection was visualized. Black arrows show the intimal flap (right).

Figure 2. Acute type A IMH (white arrow) with extension to descending aorta. Aortic diameter was 54 mm, and IMH thickness was 12 mm (left). After 6 months of medical treatment, complete regression in IMH in ascending aorta and evolution to type B dissection (black arrow) were observed (right).
dissection at a few hours or days of evolution. Interestingly, although in the series of Song et al., mortality in patients initially treated medically was low, 36.5% developed adverse clinical events within 6 months: aortic dissection in 25 (29.4%), delayed surgery in 25 (28.4%), or death in 6 (6.8%). These results coincide with those of other series in which conversion to typical dissection was reported in 30% to 40% of cases. A recent study suggested that the greatest incidence of conversion to typical dissection occurs 8 days from the onset of symptoms; however, cases of conversion occurring in the early hours or some months after the acute episode are not exceptional. Although the risk of type A IMH mortality appears to be lower than that of type A dissection, it is noteworthy that 20% of patients present with cardiac tamponade, and 30% to 40% eventually require surgical treatment owing to complications. Cardiac tamponade is an important issue in the treatment of type A IMH. Several studies reported the successful medical management with pericardiocentesis. However, when high progression rates in patients with tamponade and technical improvements in aortic surgery are considered, emergent surgery is generally preferred for this condition.

The therapeutic plan for urgent surgery for unstable patients and initial medical treatment for stable patients with surgery reserved for those who develop complications seems reasonable but requires close monitoring by imaging techniques in the early phase (Figure 2) and a prolonged hospital stay. With such an approach, a significant proportion of patients eventually require surgery during the first year of follow-up. In this respect, it is not uncommon for evolution to aortic dissection to be diagnosed during the first year of follow-up, even in the absence of symptoms. To avoid these inconveniences, it is fundamental to define the subgroup of patients with a high risk of complications. In several studies, maximum aorta diameter is the most powerful predictor of complications, with most agreeing on a diameter >50 mm as an indicator of high risk. Another imaging variable with prognostic significance, reported by Asian groups, is IMH thickness between 10 and 15 mm. Unfortunately, the lack of these data in many studies does not permit confirmation of whether the geographic variations in mortality can be explained by differences in the severity spectrum of this disease.

In conclusion, although some management strategies still remain to be elucidated, it seems clear that the mortality risk in type A IMH is lower than in classic dissection. In the subgroup of patients with hemodynamic instability, signs of impending rupture, presence of ulcer-like projections, peri-aortic hemorrhage, or tamponade, emergency treatment with surgery may be preferred. In addition, the relatively high risk of complications during IMH evolution in patients with a dilated ascending aorta and/or significant hematoma thickness would make it preferable to perform urgent surgery. When the low mortality risk of nonemergent ascending aorta surgery at expert aortic centers is considered, surgical treatment should be individualized for the rest of patients with IMH depending on variables such as the following: the option that close follow-up with serial imaging techniques is available, correct control of heart rate and blood pressure, patient age, presence of comorbidities, overall surgical expertise of the institution, and the patients’ preferences. Thus, for proximal type A IMH without the previous clinical or imaging identifiers for surgery, it is then an individualized decision between the patient and the care team. The risk of sudden unexpected deterioration and potentially preventable sudden death versus the risk of acute ascending aortic surgical repair should be weighed carefully. In a good-risk patient at a center with low surgical mortality, one might favor surgery. In a poor-risk patient at a place with high surgical mortality, medical therapy plus observation makes sense.

**Disclosures**

None.

**References**


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