Outcomes of Patients With Acute Type A Aortic Intramural Hematoma

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Background—The proper treatment option for patients with type A intramural hematoma (IMH), a variant form of classic aortic dissection (AD), remains controversial. We assessed the outcome of our institutional policy of urgent surgery for unstable patients and initial medical treatment for stable patients with surgery in cases with complications.

Methods and Results—Among 357 consecutive patients with type A acute aortic syndrome, 101 (28.3%) had IMH and 256 had AD. Urgent operations were performed in 224 patients with AD (87.5%) and 16 with unstable IMH (15.8%; P<0.001). The remaining 85 stable IMH patients received initial medical treatment, and adverse clinical events developed in 31 patients (36.5%) within 6 months, which included development of AD (n=25), delayed surgery (n=25), or death (n=6). Initial aorta diameter and hematoma thickness were independent predictors for development of these events, and the best cutoff values were 55 and 16 mm, respectively. The overall hospital mortality was lower in IMH patients than in AD patients (7.9% [8/101] versus 17.2% [44/256]; P=0.0296) and was comparable to that of surgically treated AD patients (7.9% versus 10.7% [24/224]; P=0.56). The 1-, 2-, and 3-year survival rates of IMH patients were 87.6±3.6%, 84.9±3.7%, and 83.1±4.1%, respectively. There was no statistical difference of overall survival rates between patients with IMH and surgically treated AD patients (P=0.787).

Conclusions—The clinical outcome of IMH patients receiving treatment by our policy was comparable to that of surgically treated AD patients. However, adverse clinical events were not uncommon with medical treatment alone, and initial aorta diameter and hematoma thickness may identify patients who might benefit from urgent surgery. (Circulation. 2009;120:2046-2052.)

Key Words: aorta ■ dissection ■ intramural hematoma ■ outcomes

Aortic intramural hematoma (IMH) has been increasingly recognized in patients with acute aortic syndrome and has been accepted as a unique disease entity with pathological and clinical features differing from those of classic aortic dissection (AD).1-6 Although antemortem diagnosis of IMH has become quite straightforward with the use of various noninvasive imaging techniques including contrast-enhanced computed tomography (CT), magnetic resonance imaging, and transesophageal echocardiography,2,7,8 the best approach to and treatment of IMH, especially in cases involving the ascending aorta, remain elusive. An urgent operation, as performed to treat classic proximal AD, is recommended by many investigators,3,6 but complete resorption of IMH and a favorable hospital course after medical treatment has also been reported, which suggests that aggressive medical treatment with a timed operation in selected patients showing disease progression or development of complications may be a rational treatment option.4,5,9 Much of the controversy originates from inherent limitations of previous studies and incomplete knowledge of the natural history of IMH. None of the studies performed to date has been based on a prospective randomized clinical trial, and the number of patients enrolled for each treatment (<40) was insufficient to guarantee adequate statistical power. Thus, in-depth analysis of observational data from a sizable number of patients is urgently required in an effort to characterize this relatively new disease entity and to improve our knowledge of its natural history. At our institution, initial medical treatment with timed surgery in cases with complications has been practiced for patients with stable type A IMH, and urgent surgery has been reserved for hemodynamically unstable patients; this approach is different from that of the majority of other institutions. In the present study, we sought to describe the clinical outcomes of 101 IMH patients receiving treatment by this strategy and to compare their outcomes with the primarily surgically treated AD patients.

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Methods

Subjects
From April 1993 to March 2008, 357 consecutive patients were identified prospectively at our institution as having acute aortic...
Urgent surgery was performed significantly less frequently in patients with IMH than in those with AD (15.8% [16/101] versus 87.5% [224/256]; \( P < 0.001 \)). In the AD group, except for 10 patients who refused surgery because of old age, multivariant failure at the time of clinical presentation (\( n = 10 \)) and stroke (\( n = 6 \)) were the main reasons for selecting medical treatment. Five patients died during the preparation for an urgent surgery. In the IMH group, urgent surgery was performed for patients with cardiac tamponade (\( n = 8 \)), persistent pain with suspicious progression into AD in follow-up imaging (\( n = 4 \)), and syncope at the clinical presentation (\( n = 4 \)).

**Results**

Among 357 patients, 101 (28.3%) met the strict criteria for acute proximal IMH. As shown in Table 1, patients with IMH were older than those with AD and showed a higher prevalence of female sex and hypertension. Compared with patients with AD, patients with IMH showed a greater frequency of pericardial and pleural effusion, with a higher prevalence of cardiac tamponade and syncope at clinical presentation.

Selected treatment modalities and hospital courses are summarized in Figures 1 and 2. An urgent operation was performed significantly less frequently in patients with IMH compared with those with AD (15.8% [16/101] versus 87.5% [224/256]; \( P < 0.001 \)). In the AD group, except for 10 patients who refused surgery because of old age, multivariant failure at the time of clinical presentation (\( n = 10 \)) and stroke (\( n = 6 \)) were the main reasons for selecting medical treatment. Five patients died during the preparation for an urgent surgery. In the IMH group, urgent surgery was performed for patients with cardiac tamponade (\( n = 8 \)), persistent pain with suspicious progression into AD in follow-up imaging (\( n = 4 \)), and syncope at the clinical presentation (\( n = 4 \)).

**Table 1. Demographic and Clinical Characteristics of Patients With AD and IMH**

<table>
<thead>
<tr>
<th>Variable</th>
<th>AD (n=256)</th>
<th>IMH (n=101)</th>
<th>( P )</th>
</tr>
</thead>
<tbody>
<tr>
<td>DeBakey type I/II</td>
<td>223/33</td>
<td>81/20</td>
<td>0.101</td>
</tr>
<tr>
<td>Age, y</td>
<td>56±14</td>
<td>65±10</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Male sex</td>
<td>139 (54.3)</td>
<td>30 (29.7)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Hypertension</td>
<td>134 (52.3)</td>
<td>68 (67.3)</td>
<td>0.009</td>
</tr>
<tr>
<td>Diabetes mellitus</td>
<td>7 (2.7)</td>
<td>5 (5.0)</td>
<td>0.366</td>
</tr>
<tr>
<td>Marfan syndrome</td>
<td>20 (7.8)</td>
<td>1 (1.0)</td>
<td>0.011</td>
</tr>
<tr>
<td>Bicuspid aortic valve</td>
<td>4 (1.6)</td>
<td>1 (1.0)</td>
<td>1.000</td>
</tr>
<tr>
<td>Smoking</td>
<td>52 (47.3)</td>
<td>16 (32.0)</td>
<td>0.227</td>
</tr>
<tr>
<td>Maximal aortic diameter, mm</td>
<td>52.1±8.4</td>
<td>50.1±8.1</td>
<td>0.052</td>
</tr>
<tr>
<td>Pericardial effusion</td>
<td>122 (47.7)</td>
<td>69 (68.3)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Pleural effusion</td>
<td>61 (24.5)</td>
<td>49 (49.0)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Cardiac tamponade</td>
<td>24 (9.4)</td>
<td>21 (20.8)</td>
<td>0.004</td>
</tr>
<tr>
<td>Syncope</td>
<td>22 (8.6)</td>
<td>19 (18.8)</td>
<td>0.009</td>
</tr>
<tr>
<td>Azotemia</td>
<td>83 (32.8)</td>
<td>24 (24.2)</td>
<td>0.124</td>
</tr>
<tr>
<td>(creatinine &gt;1.4 mg/dL)</td>
<td>10 (3.9)</td>
<td>2 (2.0)</td>
<td>0.521</td>
</tr>
<tr>
<td>Focal neurological sign</td>
<td>4 (1.6)</td>
<td>0 (0.0)</td>
<td>0.581</td>
</tr>
<tr>
<td>Visceral ischemia</td>
<td>106 (45.9)</td>
<td>12 (12.1)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

**Note:** Values in parentheses are percentages.

syndrome involving the ascending aorta. Patients were diagnosed by routine clinical evaluation consisting of random combinations of CT, magnetic resonance imaging, and transesophageal echocardiography; >80% of patients underwent both CT and echocardiography. Only patients who visited the hospital within 14 days of symptom onset were included in the present study. Such patients were analyzed and separated into those with classic AD and those with typical IMH. A classic double-channel aorta with a visible intimal tear or flap was a diagnostic criterion for AD, and a crescentic or circular high-attenuation area along the aortic wall, seen without contrast enhancement on CT, and regional thickening of the aortic wall on echocardiography, without evidence of flow communication through the intimal flap, were considered diagnostic of IMH. Any patient with a penetrating aortic ulcer at the aortic arch and localized IMH adjacent to the ulcer was excluded from analysis in this study. We have published clinical data of patients admitted until March 2003 elsewhere, and the number of the subjects in this study almost doubles those of the last report. In our institution, from April 1999, initial medical treatment with timed surgical intervention in cases with complications on follow-up imaging study has been recommended for type A IMH. Urgent surgery was recommended for hemodynamically unstable patients.

**Data Collection and Analysis**

Chart review was performed, and data were collected with the use of a standardized form that recorded information on patient demographics, medical history, clinical presentation, physical findings, results of imaging studies, details of medical and surgical treatment, and outcome. An urgent operation was defined as surgery performed within 3 days of hospital admission. Patients with IMH were compared with those with classic AD with respect to demographic findings, presentation, treatment, and outcome.

Follow-up data were collected by direct telephone interview and a detailed review of all medical records. The cause and date of any death were confirmed by information gathered from the National Population Registry of the Korean National Statistical Office, together with a review of all available clinical records at the time of death. Adverse clinical events included development of AD, delayed surgery, and cardiovascular death. Mean follow-up duration was 3.1±3.3 years (median, 2.0 years; range, 0 to 14.0 years).

**Statistical Analysis**

All statistical analyses were performed with the use of SPSS (version 12.0; SPSS Inc, Chicago, Ill). Summary statistics are presented as frequencies and percentages, mean±SD values, or medians with interquartile ranges. Statistical analysis of differences between groups for continuous variables was assessed by means of means of Student \( t \) tests. The \( \chi^2 \) tests and Fisher exact tests were used to compare the frequency between groups. Cumulative survival rate curves were generated with the Kaplan–Meier method and compared with the log-rank test. Univariable and multivariable Cox proportional hazard models were used to determine factors associated with development of adverse clinical events (death, development of classic AD, or delayed surgery) during medical treatment in patients with IMH. Multivariable analyses involved a backward elimination technique, and only variables with a \( P \) value of <0.10 were used in the final model. To assess the cutoff point of initial hematoma thickness and aorta diameter for predicting development of adverse clinical events with medical treatment, receiver operating characteristic curves were used. The optimal cutoff value was defined as the value with the maximal sum of sensitivity and specificity. All \( P \) values were 2-sided, and a value of \( P<0.05 \) was considered significant.

**Figure 1. Outcomes of patients with type A AD according to initial treatment option.**
Medical treatment included intravenous injection of \( \beta \)-adrenergic receptor blocker accompanied by a long-acting calcium channel blocker to maintain the systolic blood pressure \( \geq 120 \) mm Hg during the acute stage. Follow-up CT or transesophageal echocardiography was performed every week or when a patient complained of renewed pain. Among 85 patients who received initial medical treatment, delayed surgery was performed in 25 (29.4%); a timed operation during hospital admission was done in 17; and surgery was performed during clinical follow-up after uneventful discharge in an additional 8 patients. Development of classic AD was the most common reason for surgical intervention \( (n = 16) \), whereas other causes of surgery included increase in IMH thickness \( (n = 5) \) or aortic dimension \( (n = 1) \) or no change in hematoma thickness \( (n = 1) \). In 2 patients who showed no change of IMH thickness and no evidence of AD in follow-up imaging studies, an elective operation was done because of the presence of Marfan syndrome \( (n = 1) \) and a progressive increase of pleural effusion \( (n = 1) \). The median duration from symptom onset to delayed surgery in these 25 patients was 27 days (interquartile range, 17 to 38 days), and the mortality rate was 4% \( (1/25) \).

The overall hospital mortality was lower in IMH patients than in AD patients \( (7.9\% \ [8/101] \ versus \ 17.2\% \ [44/256] ; P=0.0296) \) and was comparable to that of surgically treated AD patients \( (7.9\% \ versus \ 10.7\% \ [24/224] ; P=0.56) \). The hospital mortality of patients with AD who received medical treatment was higher than those who underwent an urgent operation \( (62.5\% \ [20/32] \ versus \ 10.7\% \ [24/224] ; P<0.0001) \), whereas mortality in the IMH group did not differ statistically between those who did and did not undergo an urgent operation \( (12.5\% \ [2/16] \ versus \ 7.1\% \ [6/85] ; P=0.609) \). The clinical profiles of 6 patients with IMH who died without emergent surgical intervention during the acute stage are summarized in Table 2. In 2 patients, an operation was contraindicated because of stroke (case 1) and old age (case 2). One patient suffered sudden death in the emergency department as the patient awaited hospital admission (case 4), and another patient died before CT (case 3). Case 5 showed development of AD in short-term follow-up imaging and suddenly died awaiting emergent surgery. Case 6 died suddenly in the intensive care unit without complaining of any chest discomfort or pain.

After hospital discharge, cardiovascular problems and sudden death developed in 4 patients in the medically treated

### Table 2. Clinical Data on Patients With IMH Who Died During Hospital Management or Follow-Up

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age, y/Sex</th>
<th>Diameter/Thickness, mm</th>
<th>Emergent Surgery</th>
<th>Hospital Course and Outcome</th>
<th>Time to Death</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>70/M</td>
<td>67/42</td>
<td>No</td>
<td>Presented with stroke and died of increased intracranial pressure</td>
<td>2 d</td>
</tr>
<tr>
<td>2</td>
<td>91/F</td>
<td>55/10</td>
<td>No</td>
<td>Tamponade (+); refused surgery after pericardiocentesis</td>
<td>1 d</td>
</tr>
<tr>
<td>3</td>
<td>64/F</td>
<td>60/17</td>
<td>No</td>
<td>Sudden death waiting for CT</td>
<td>1 d</td>
</tr>
<tr>
<td>4</td>
<td>69/F</td>
<td>52/19</td>
<td>No</td>
<td>Sudden death waiting for hospital admission</td>
<td>1 d</td>
</tr>
<tr>
<td>5</td>
<td>41/M</td>
<td>57/28</td>
<td>No</td>
<td>AD confirmed 2 days later; sudden death waiting for emergent surgery</td>
<td>3 d</td>
</tr>
<tr>
<td>6</td>
<td>71/F</td>
<td>60/16</td>
<td>No</td>
<td>Sudden death during hospital admission</td>
<td>5 d</td>
</tr>
<tr>
<td>7</td>
<td>78/F</td>
<td>60/20</td>
<td>No</td>
<td>Discharged alive; AD confirmed 2 months later; contraindication to surgery because of severe asthma</td>
<td>17.5 mo</td>
</tr>
<tr>
<td>8</td>
<td>74/F</td>
<td>56/16</td>
<td>No</td>
<td>Discharged alive; AD confirmed 1 month later; died after surgery of postoperative sepsis</td>
<td>2.0 mo</td>
</tr>
<tr>
<td>9</td>
<td>71/F</td>
<td>50/10</td>
<td>No</td>
<td>Discharged alive; died suddenly during follow-up; no evidence of development of AD before death</td>
<td>35.8 mo</td>
</tr>
<tr>
<td>10</td>
<td>77/F</td>
<td>47/12</td>
<td>No</td>
<td>Discharged alive; died suddenly during follow-up; no evidence of development of AD before death</td>
<td>5.8 mo</td>
</tr>
<tr>
<td>11</td>
<td>72/F</td>
<td>Yes</td>
<td></td>
<td>Died of postoperative pneumonia</td>
<td>10 d</td>
</tr>
<tr>
<td>12</td>
<td>68/F</td>
<td>Yes</td>
<td></td>
<td>Died of postoperative bleeding</td>
<td>1 d</td>
</tr>
<tr>
<td>13</td>
<td>76/F</td>
<td>Yes</td>
<td></td>
<td>Discharged alive; died suddenly during follow-up; no evidence of development of AD before death</td>
<td>5.0 mo</td>
</tr>
<tr>
<td>14</td>
<td>62/F</td>
<td>Yes</td>
<td></td>
<td>Discharged alive; died suddenly during follow-up; no evidence of development of AD before death</td>
<td>13.7 mo</td>
</tr>
</tbody>
</table>

Diameter/thickness indicates maximal aortic diameter/maximal hematoma thickness.
group and in 2 patients after surgery. In 2 patients in the medically treated group (cases 7 and 8), development of classic AD was confirmed by follow-up imaging. Surgery was contraindicated in 1 patient because of severe asthma and poor results of pulmonary function test, and this patient died (case 7). The other patient underwent surgery and died of sepsis associated with the surgical wound (case 8). The other 2 patients died suddenly during follow-up (cases 9 and 10). In the urgent surgery group, operative mortality developed in 2 patients (cases 11 and 12), and 2 patients died suddenly during follow-up (cases 13 and 14). The 1-, 2-, and 3-year survival rates of our patients with IMH were 87.6 ± 3.6%, 84.9 ± 3.7%, and 83.1 ± 4.1%, respectively. There was no statistical difference in overall survival rates between patients with IMH and surgically treated AD patients (Figure 3). The 3-year survival rate of stable IMH patients with initial medical treatment was 86.3 ± 4.2%.

Among 85 stable IMH patients who received initial medical treatment, 31 (36.5%) experienced adverse clinical events including development of AD (n=25), delayed surgery (n=25), or death (n=6) within 6 months. Patients who developed adverse clinical events after medical treatment showed a higher prevalence of syncope (29.0% versus 11.1%) with a larger aortic diameter (53.6 ± 6.6 versus 47.5 ± 8.2 mm) and a thicker hematoma in the initial imaging study on clinical presentation (14.6 ± 7.8 versus 11.0 ± 7.6 mm). In multivariable analysis with the Cox proportional hazard model, syncope (hazard ratio=3.534; 95% confidence interval [CI], 1.527 to 8.183; P=0.003), hematoma thickness (hazard ratio per millimeter=1.076; 95% CI, 1.018 to 1.137; P=0.010), and aorta diameter (hazard ratio per millimeter=1.066; 95% CI, 1.006 to 1.128; P=0.029) were independent predictors for development of adverse clinical events after medical treatment. The best cutoff values of hematoma thickness and aorta diameter were 16 mm (area under the curve=0.70; 95% CI, 0.58 to 0.81; P<0.001) and 55 mm (area under the curve=0.73; 95% CI, 0.62 to 0.84; P<0.001), respectively. The event-free survival rate was significantly different depending on whether a patient had neither, either, or both risk factors (Figure 4).

In our study, it was possible to analyze the temporal pattern of AD development during medical treatment of IMH patients. As shown in Figure 5, AD developed within 1 month after symptom onset in >60% (16/25) of the affected patients. However, development of AD could occur at any time during medical treatment. Three patients died after confirmation of AD development; 1 patient (case 5) died suddenly, while awaiting surgery, in the hyperacute stage (2 days after symptom onset), and another patient (case 8) who developed AD 1 month after initial symptom onset could not undergo surgery because of intractable asthma and died 1.5 years after symptom onset of aorta rupture. The other patient (case 7) who developed AD 2 months after initial symptom presentation died of postoperative sepsis.

**Discussion**

In this observational study, we found that patients with IMH constituted a significant proportion of type A acute aortic syndrome patients and that these 2 classes of patients showed different clinical features and demonstrated a more favorable
response to medical treatment than did patients with classic AD. We also found that although development of classic AD and sudden death occurred not infrequently during medical treatment, unlike in AD patients, our policy of urgent surgery for unstable type A IMH patients and initial medical treatment with close follow-up for patients with stable type A IMH showed acceptable clinical outcomes. Initial hematoma thickness and aortic diameter provided important prognostic information, which could be used for risk stratification and clinical decision making with respect to treatment strategy and surgical timing.

**Type A IMH: Clinical Features and Natural History With Medical Treatment**

The prevalence of type A IMH in our study was 28.3% (101/357), which is much higher than that (3.6% [23/639]) recently reported in the largest multicenter registry currently available (the International Registry of Aortic Dissection [IRAD]), but is comparable to the prevalence reported by other Japanese investigators. Although prominent geographic or racial differences in the relative prevalence of IMH may exist between Eastern and Western countries, it is interesting to observe the universal finding that the relative incidence of IMH affecting the distal aorta is much higher than that affecting the proximal aorta. The prevalence values of type A and B IMH in IRAD are 3.6% and 9.4% (35/371), respectively. According to the Japanese and Korean studies, the relative prevalence of type B IMH (48.2% [53/110] to 58.3% [108/185]) was >2-fold higher than that of type A IMH (22.9% [30/131] to 27.4% [51/186]). Thus, unlike the situation with AD, which more frequently affects the ascending aorta, IMH has a strong tendency to involve the distal aorta.

Patients with IMH were older than those with AD. This is another universal finding reported by previous investigators, regardless of geographic location. The almost 1-decade difference in mean age between AD and IMH patients, as well as the markedly different prevalence of Marfan syndrome and hypertension, might indicate that different pathophysiological mechanisms are responsible for the development of AD and IMH. Besides the possibility of higher incidence of death before diagnosis in patients with AD and tamponade, the higher frequency of pericardial and pleural effusion in IMH patients could be explained partly by the more eccentric location of IMH (toward the adventitia), associated with an increased tendency to extravasation.

Apart from international heterogeneity in the relative prevalence of type A IMH, geographic difference in clinical outcome with medical treatment and a role for urgent surgical intervention remain challenging issues. European investigators have reported very high mortality rates after medical treatment alone and have advocated urgent surgery, whereas studies in Japan and Korea showed comparable outcomes in the absence of urgent surgery. These studies were not randomized trials, and enrolled patient numbers were <40 even in multicenter studies; these numbers are certainly too low to offer the required statistical power. In the present study, despite unfavorable conditions of higher mean age and significantly lower frequency of urgent surgery in IMH patients, hospital mortality and survival rates were comparable to those of surgically treated AD patients. These findings represent a more favorable response to medical treatment of IMH patients, possibly because of the absence of continuous flow communication through an intimal tear in IMH. The low overall hospital mortality (7.9%) in our study differs from the results of a recent IRAD report in which the overall mortality of type A IMH patients was 39.1% (9/23) and mortality rates associated with medical and surgical treatment were 33.3% (3/9) and 42.9% (6/14), respectively. The observed ascertainment bias of the IRAD registry and different indications for urgent surgery between IRAD report and our study might explain the mortality difference. However, it is interesting to find that, even in the IRAD report, there was no mortality difference between patients receiving medical or surgical treatment for type A IMH (33.3% versus 42.9%; P=1.00), and type A IMH patients were less likely to receive surgery compared with patients with AD (60.9% versus 82.5%; P=0.023).

The IRAD report distinguished between medical and surgical treatment for type A IMH but did not stratify between urgent and nonurgent surgery. If we reanalyze our data as the IRAD investigators did, 33 patients (32.7%), including 17 who received elective surgery during hospital admission, could be classified as being in the surgery group, whereas the remaining 68 could be classified as being in the medical group. The mortality did not differ statistically between the groups (6.1% [2/33] versus 8.8% [6/68]; P=1.0). According to a recent meta-analysis of type A IMH, there was no statistically significant difference in mortality rate between the initial surgery and medical group (10.1% [17/168] versus 14.4% [23/160]; P=0.37), which is consistent with our results.

**Adverse Events Without Urgent Surgery and Prognostic Role of Initial Imaging Study**

Although we have shown that our policy of urgent surgery for unstable type A IMH patients and initial medical treatment for stable patients with surgery in cases with complications had acceptable outcomes, and although dramatic resorption of hematoma in the hyperacute phase without emergent surgery has been reported, occurrence of sudden death or AD still remains a potential threat in patients receiving initial medical treatment. Moreover, a delayed operation in the initial medical treatment group was not infrequently needed; the operative frequency was 32% (25/79) in our study, which is intermediate between the previously reported frequencies of 27% (7/26) and 43% (13/30). These findings reinforce the importance of thorough analysis to characterize the clinical features of any adverse event and to seek valuable predictors of such occurrences.

Sudden death is the most tragic event associated with IMH, and the most probable reason for sudden death is aortic rupture. Early investigators already reported that aortic rupture occurred much more commonly in IMH patients than in those with type A or B AD, resulting in high mortality. In our study, among 6 patients who died in the acute stage during management without urgent surgery, 4 (cases 3 to 6, 66.7%) died unexpectedly within 5 days of symptom onset.
Among these, development of typical AD could be demonstrated in only 1 patient (case 5). Sudden death resulting from aortic rupture is believed to represent increased permeability of the aortic wall, and development of classic AD seems not to be a prerequisite for sudden death.\textsuperscript{2,14,20} In other studies, all aortic ruptures were reported to occur on initial presentation or during initial hospitalization\textsuperscript{6,20}; however, in our study, sudden death also occurred during follow-up after an eventful medical treatment (n=2 [cases 9 and 10]) or successful surgery (n=2 [cases 13 and 14]). It is not certain whether sudden death during long-term clinical follow-up was directly related to IMH in these 4 patients because autopsies were not performed.

Development of typical AD has been described as another ominous adverse clinical event in IMH patients and is reported to be a major reason for a timed or delayed operation.\textsuperscript{5,19} However, the temporal pattern and its prognostic impact have not been investigated in detail. In the present study, AD developed in a rather random fashion within 2 months from initial symptom onset, and the risk of AD development seems not to be critically time dependent. Thus, physicians should be aware that typical AD can occur at any time during the course of initial medical treatment. One important finding is that development of AD is not necessarily associated with sudden death. Because development of typical AD is accompanied by vague chest discomfort or pain, physicians need to be vigilant in efforts to detect this potentially fatal complication.

The prognostic role of imaging studies for risk stratification cannot be overemphasized.\textsuperscript{21} As found in previous studies with fewer patients,\textsuperscript{9,22–24} we have reconfirmed that initial maximal aortic dimension and hematoma thickness have strong prognostic power to predict the development of adverse clinical events, including death in the acute phase, AD development, or a need for timed surgery during initial medical management. Although our cutoff values were not validated in an independent data set, it is interesting to observe that all 5 patients who died suddenly during the acute stage showed maximal hematoma thickness equal to or greater than the cutoff value (16 mm), and urgent surgery based on this finding might be helpful to prevent sudden death. Thus, noninvasive imaging techniques provide not only an accurate differential diagnosis of acute aortic syndrome but also prognostic information useful for risk stratification.\textsuperscript{21} We believe that clinical decision making based on these measurements is rational and that the initial imaging study may identify hemodynamically stable patients who might benefit from urgent surgery. Recommendation of emergent surgery in stable patients with a very thick hematoma or a large aorta might reduce the risk of sudden death or hospital mortality; further clinical investigation is necessary to test this hypothesis.

Limitations

Although our data are based on the largest numbers of patients with type A IMH examined to date, this is an observational study in a single center, and our results may not apply to European and North American populations. On the basis of the significantly lower relative risk of surgical treatment in a meta-analysis,\textsuperscript{22} most surgeons still advocate urgent surgery to treat type A IMH, as used in classic AD.\textsuperscript{26,27} Meta-analysis also has a number of limitations, including publication and reporting bias. The most difficult point in the interpretation of the meta-analysis or multicenter registry data is that the description of IMH thickness or maximal aortic diameter is incomplete, and comparison of IMH severity is therefore not possible. Because a major difference, caused by selection bias, in the clinical features and outcomes of patients with penetrating aortic ulcers has been demonstrated in 2 recent publications,\textsuperscript{26,27} variations between our data and those of previous reports might be explained by different levels of IMH severity.\textsuperscript{30}

Conclusions

In this study population, acute IMH accounts for 28% of acute aortic syndrome involving the ascending aorta. Clinical features differ from those of classic AD, and overall mortality is lower compared with that seen in patients with AD. Unlike in AD patients, our policy of urgent surgery for unstable type A IMH patients and initial medical treatment for stable patients with surgery for complications showed acceptable outcomes. However, fatal aortic rupture may occur in the hyperacute stage, and progression to classic AD is not infrequent and requires a delayed operation. Initial aortic diameter and hematoma thickness, reflecting IMH severity, are excellent predictors of development of these adverse clinical events in the absence of urgent surgery, and surgical intervention should be considered in patients with these risk factors. Thus, we believe that the spectrum of IMH severity is broad, with diverse clinical outcomes, and further investigations are needed to conclude whether risk stratification and selection of an appropriate treatment option based on an initial imaging study are both feasible and safe.

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Disclosures

None.

References

Aortic intramural hematoma (IMH) is increasingly recognized as a unique disease entity with pathological and clinical features differing from those of classic aortic dissection. However, the best treatment of IMH, especially in cases involving the ascending aorta, remains uncertain. We assessed the outcomes of our institutional policy of urgent surgery for unstable type A IMH patients and initial medical treatment for stable patients with surgery for subsequent complications. Type A IMH occurred in 101 patients (28%) admitted with a type A acute aortic syndrome. IMH patients were older than those with type A IMH patients and initial medical treatment for stable patients with surgery for subsequent complications. Type A IMH is a unique disease entity with more favorable short-term evolution of aortic intramural hematoma: therapeutic implications. Eur Heart J. 2007;28:1158–1163.


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