Clinical Features and Prognosis in Dissecting Aneurysm of the Aorta

A Re-appraisal

By Joseph Lindsay, Jr., M.D., and J. Willis Hurst, M.D.

SUMMARY

The records of 62 patients on whom the diagnosis of aortic dissection was proved are reviewed with particular attention to the clinical features and prognosis in each case. This was the consecutive experience of a large general hospital over approximately 17 years. The initial and long-term survival was far better in patients in whom the ascending aorta was spared by the disease process. No patients in whom such involvement was present were known to survive more than 3 weeks. Eight of 19 patients whose disease began distal to the arch of the aorta are known to have survived 6 to 69 months even though six of these eight were not operated on. Striking differences in the clinical findings of the two groups were also found. Aortic regurgitation, impairment of a radial or carotid pulse, neurological signs, and hypotension were rare in the group whose dissection began beyond the left subclavian artery. Marked systemic hypertension was frequently observed in this group, over a third having a diastolic pressure over 140 mm Hg and nearly two thirds over 120 mm Hg. Such hypertension was unusual when the ascending aorta was the origin of the disease. Despite the present study the available data do not allow any definite conclusions about the efficacy of any form of therapy of this disorder.

Additional Indexing Words:
Hypertension  Pain
Race  Neurological findings
Aortic regurgitation

The prognosis of patients with dissecting aortic aneurysm is regarded as poor. In a review of 425 cases, Hirst and associates found a 37% mortality in the first 48 hours. Only 26% of their patients survived 2 weeks and less than 10% were alive for 1 year. Surgical attack on this lesion entails a very high risk, particularly when the dissection begins in the ascending aorta. Despite this, operation is usually recommended because of the poor prognosis in the untreated case.

Recently, reports have appeared which suggest that the natural history of this disorder may be incompletely understood. Hume and Porter pointed out that the present information regarding the course of aortic dissection is based on large series of autopsied cases which have been collected from the literature. These authors feel that such series are subject to various biases and that the analysis of a large number of consecutive cases might provide a more realistic picture of the expected course of this disease.

The need for an understanding of the natural history of aortic dissection is now even more imperative. This is true because Wheat and associates have reported the survival without surgery of 10 consecutive patients with acute dissecting aortic aneurysm. They employed vigorous drug therapy directed toward lowering the arterial pressure and
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thereby the left ventricular “impulse.” This alternative to surgical therapy must be critically evaluated. It seemed desirable, therefore, to review a consecutive, unselected series of patients who received primary treatment in a large general hospital. A by-product of the study was the opportunity for reappraisal of the clinical features of this disease.

Patients Studied

All records indexed as dissecting aortic aneurysm at Grady Memorial Hospital from January 1949 through February 1966 were reviewed. Those cases in which there was autopsy, surgical, or aortographic proof of the diagnosis were accepted for the study group. Dissecting aneurysms not involving the thoracic aorta were excluded. No patient had been hospitalized elsewhere prior to his initial contact with this hospital. The records of 62 patients met these criteria and were further studied in detail and appropriate notations of the clinical and laboratory findings were made. Approximately 30 records, in which a diagnosis of aortic dissection was not verified, were excluded from this study group.

Results

Extent of the Dissecting Process

The distribution of involvement of the aorta by the dissecting process in the present series is shown in table 1. The extent of the dissection was determined from the autopsy protocol, the surgical record, or the findings on retrograde aortography.

The ascending aorta was involved in 40 patients (65%). The process began in the aorta distal to the arch in 19 patients (31%). Twenty-nine instances (47%) of end-to-end aortic dissection were observed.

Thirty-eight of our cases (61%) may clearly be labeled as cases of dissection, type I by the criteria of DeBakey and associates for the dissection began in the ascending aorta and extended distally. It seems appropriate to include as type I also the two cases in which the process was limited to the ascending aorta since neither had stigmata of Marfan’s syndrome and both had an acute course.

<table>
<thead>
<tr>
<th>Extent of Involvement of Aorta by Dissection</th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ascending aorta to iliac arteries</td>
<td>29</td>
</tr>
<tr>
<td>Ascending aorta into abdomen</td>
<td>8</td>
</tr>
<tr>
<td>Ascending aorta to descending abdomen</td>
<td>1</td>
</tr>
<tr>
<td>Limited to ascending aorta</td>
<td>2</td>
</tr>
<tr>
<td>Limited to arch of the aorta</td>
<td>1</td>
</tr>
<tr>
<td>Distal to left subclavian artery</td>
<td>19</td>
</tr>
<tr>
<td>Undetermined due to insufficient data</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>62</td>
</tr>
</tbody>
</table>

The 19 cases (31%) in which dissection began distal to the left subclavian artery may be classified type III, of DeBakey, and will be so considered in the discussion to follow.

Three patients could not be easily fitted into the DeBakey classification. One was found to have a 6-cm dissection within the arch which extended into all the major arch vessels. The records of two other patients did not give the extent of the dissection.

Comment

Aortic dissection began in the ascending aorta in more than half of the patients reported in most large series. In another quarter of these cases, the process began distal to the left subclavian artery. Distal extension, usually below the diaphragm and often to the iliac arteries, was the rule. Proximal extension into the ascending aorta from those dissections which began distal to the arch vessels occurred but was more unusual.

DeBakey and associates classified aortic dissection from a surgical standpoint into three general types based upon the origin and extent. Those of type I characteristically begin in the ascending aorta and extend distally for a variable distance. The entire length of the aorta is often involved in type I dissection. Type II includes those patients in whom the process is confined to the ascending aorta. DeBakey’s patients in this group all had stigmata of Marfan’s syndrome and all were seen in the chronic stage of their illness. Those instances in which the disease begins distal to the arch vessels make up type III.

Circulation, Volume XXXV, May 1967
Clinical Features

Age and Sex

Age and sex distribution in the present group of patients is comparable to that reported by other authors. Males predominate with a ratio of 40 to 22 females.

In 40 instances, symptoms of dissection began in the sixth or seventh decade. The youngest patient was 31 and the oldest 83 years of age. Age distribution by decades is shown in Table 2.

Fifty-two of the patients (84%) were Negroes. The remainder were Caucasian. Eighty-four per cent exceeds the percentage of Negro admissions to this hospital and suggests a relatively increased prevalence of aortic dissection in that race. It may be conjectured that a relationship exists to the increased frequency of hypertensive vascular disease reported in the Negro race.

Hypertension

Fifty patients (81%) were either hypertensive on admission or gave a history of systemic hypertension in the past. There was no difference between Negroes and Caucasians in the prevalence of these factors in the current study. Few individuals were under medical supervision for high blood pressure; two patients, however, were receiving guanethidine and another was being given mecamylamine at the time dissection occurred. Good control of their blood pressure had not been achieved.

Table 2

<table>
<thead>
<tr>
<th>Age Distribution of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (yr)</td>
</tr>
<tr>
<td>30 to 39</td>
</tr>
<tr>
<td>40 to 49</td>
</tr>
<tr>
<td>50 to 59</td>
</tr>
<tr>
<td>60 to 69</td>
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<tr>
<td>70 to 79</td>
</tr>
<tr>
<td>80 to 89</td>
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<tr>
<td>Total</td>
</tr>
</tbody>
</table>

* Aortic dissection involving the ascending aorta.
† Aortic dissection beginning distal to the arch vessels.

Pain

Pain, always a common symptom of aortic dissection, was the presenting symptom in 85% in the series of Hirst and associates. Fifty-six patients (90%) in the present group gave a history of pain which could be reasonably ascribed to this process. Of the remaining six, five had severe neurological defects on admission and a history of discomfort could not have been obtained. The remaining patient died suddenly in the hospital without ever complaining of pain.

The initial sites of pain are tabulated in Table 3. Burchell reported that the location of the pain was helpful in determining the location of the dissecting process, but this was not true in our patients.

Table 3

<table>
<thead>
<tr>
<th>Site of Principal Pain</th>
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</thead>
<tbody>
<tr>
<td>Site</td>
</tr>
<tr>
<td>Anterior chest</td>
</tr>
<tr>
<td>Interscapular area</td>
</tr>
<tr>
<td>Epigastrium</td>
</tr>
<tr>
<td>Low back</td>
</tr>
<tr>
<td>Extremity</td>
</tr>
<tr>
<td>No pain</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>

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DISSECTING ANEURYSM OF THE AORTA

Two patients entered the hospital because of altered consciousness of varying degrees of 3 weeks’ and 18 months’ duration. No history of pain was obtained and the aortic dissection was a surprise finding at autopsy in both.

The acute onset of hemiparesis was noted in four cases and paraparesis in one. The patient with 3 weeks of mental obtundation also had a mild hemiparesis for that period of time.

Ischemic paralysis of a leg was noted in nine instances.

One patient whose dissection was limited to the descending aorta was confused, but no other neurological findings were recorded during the acute episode in patients with this form of the disease.

The neurological findings occurring in the acute episode are shown in table 4.

Aortic Regurgitation

The murmur of aortic regurgitation was audible on admission in 22 of the patients (35%) in the present series. A pre-existing diastolic murmur was known in one patient, and an aortic valve deformity compatible with rheumatic heart disease was observed at autopsy in a second. No examination was recorded prior to his admission with aortic dissection. In two additional patients, whose dissection was shown by aortography to be limited to the descending aorta, the murmur appeared 2 or more weeks after the initial insult. This may have been due to retrograde extension of the process; however, these two patients are still alive and such extension has not been demonstrated.

Alterations in Pulses

Detectable alterations in the peripheral arterial pulses were recorded in 28 patients (45%). One or more of the carotid or upper extremity pulses was altered in 14 subjects.

Pulse alteration was found in only two patients with type III dissection. In one instance, both the right carotid and the right femoral pulses were altered; in another instance the right femoral pulse was reduced. The patient whose right carotid pulse seemed altered was one of those in whom aortic regurgitation was a late finding.

Clinical Findings Related to Extent of Dissection

We expected that aortic dissection involving the ascending aorta would often produce aortic regurgitation and its extension distally might compromise pulses of the arch vessels. We did not expect dissection limited to the descending aorta to produce such findings. The relationship of certain clinical findings to the distribution of the process is shown in figure 1.

As expected 16 of 19 patients with type III dissection had neither aortic regurgitation

<table>
<thead>
<tr>
<th>Neurological Manifestations Occurring Acutely</th>
</tr>
</thead>
<tbody>
<tr>
<td>Manifestation</td>
</tr>
<tr>
<td>----------------------------</td>
</tr>
<tr>
<td>Altered consciousness</td>
</tr>
<tr>
<td>Hemiparesis</td>
</tr>
<tr>
<td>Paraparesis</td>
</tr>
<tr>
<td>Ischemic paralysis</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>

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Figure 1

Frequency of certain physical findings in patients with aortic dissection. The frequency of an aortic regurgitant murmur and of the alteration of certain pulses in type I patients is contrasted with the relative infrequency of these findings in type III patients. The 40 patients in whom the ascending aorta was involved are labeled type I. The 19 patients in whom the dissection began distal to the arch vessels are labeled type III.
nor compromise of an arch vessel on admission. The murmur of aortic regurgitation occurred subsequently in two patients from this group, one of whom had had a diminished right carotid pulse on admission. Fifteen patients (79%) with type III and five patients (13%) with type I dissections had neither aortic regurgitation nor a pulse deficit of any sort.

Marked systemic hypertension was a common clinical finding in the patients whose dissection began distal to the arch vessels. Eight (42%) had a diastolic pressure of 140 mm Hg or greater on admission and in four of these, diastolic pressure levels of 160 to 170 mm Hg were recorded. Figure 2 emphasizes that such pressures were unusual in patients with type I involvement of the aorta. Frank shock levels of blood pressure were not observed in patients with type III dissection but were found in 20% of those with type I. We did not foresee these distinctive differences in the two groups of patients.

It would appear that the majority of patients whose lesion begins in the ascending aorta present with a clue that such involvement is present. However, the absence of such clues does not guarantee that the process is limited to the descending aorta since 13 of our patients (33%) with involvement of ascending aorta had neither aortic regurgitant murmurs nor detectable involvement of an arch vessel. In contrast most of the patients who had type III dissection presented with pain as the only suggestive clinical manifestation although marked hypertension was often present.

**Prognosis**

Length of survival was determined from the onset of symptoms of aortic dissection. There was usually characteristic pain although the sudden loss of consciousness marked the onset in a few instances. In four instances, no history characteristic of dissection was obtained.

Mortality within the first month is shown in figure 3. Those instances in which the duration of survival was uncertain are considered to have lived at least 1 month.

![Figure 2](image)

**Figure 2**

Frequency of marked hypertension or of hypotension in patients with aortic dissection. The increased prevalence of marked hypertension in type III patients (dissection distal to arch vessels) when compared with patients in type I (dissection in ascending aorta) is depicted. Severe hypotension occurred only in patients with involvement of the ascending aorta.

![Figure 3](image)

**Figure 3**

Survival during the first month of illness. The survival during the first month of illness of patients in the two groups is contrasted. Patients in whom the duration of survival was uncertain are considered to have survived longer than 1 month. Type III = dissection distal to arch vessels; type I = dissection of ascending aorta.
Early mortality in the present series is similar to that reported by most large series in the literature. It is apparent that the early prognosis for patients whose dissection is limited to the descending aorta is far better than the early prognosis for the other group.

Among the 40 patients whose dissection involved the ascending aorta, the duration of survival was uncertain in two and may have been as long as several months. None of the remainder survived their initial hospitalization. Longest known survival was 3 weeks even though eight underwent emergency fenestration procedures.

External rupture of the aorta was the cause of death in 31 patients. Hemopericardium was present in each of these. Bilateral hemothorax was an additional finding in three and hemomediatinum was present in three. Death was attributable to the dissecting process in all instances. Table 5 lists the cause of death in this group of patients.

Table 5

<table>
<thead>
<tr>
<th>Cause of death</th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>External rupture</td>
<td>31</td>
</tr>
<tr>
<td>Hemopericardium</td>
<td>31</td>
</tr>
<tr>
<td>Hemothorax (bilateral)</td>
<td>3</td>
</tr>
<tr>
<td>Hemomediatinum</td>
<td>3</td>
</tr>
<tr>
<td>Cardiac arrest at surgery</td>
<td>2</td>
</tr>
<tr>
<td>Shock</td>
<td>2</td>
</tr>
<tr>
<td>Encephalomalacia and pneumonia</td>
<td>1</td>
</tr>
<tr>
<td>Congestive heart failure</td>
<td>1</td>
</tr>
<tr>
<td>Myocardial infarction (coronary occlusion by the dissection)</td>
<td>1</td>
</tr>
<tr>
<td>Uncertain (sudden death in first 24 hours)</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>40</td>
</tr>
</tbody>
</table>

The onset of the disease in one patient whose disease began distal to the arch vessels could not be ascertained. Twelve of the remaining 18 patients with type III dissection were not operated on during their initial hospitalization. Table 6 considers the course of these patients.

Five of the six patients who died without surgery during their initial hospitalization did so because of external rupture. One other died of extensive pneumonia. In three of the four patients who died after they had left the hospital, autopsy revealed cerebral vascular accidents unrelated to aortic dissection. One patient died on the operating table 5 years and 9 months after her original dissection while an attempt was being made to replace the descending thoracic portion of her aorta. One patient died of hypertensive encephalopathy. Old aortic dissection was a surprise autopsy finding. All four of the postoperative deaths were due to surgical complications. Table 7 summarizes the cause of death in this group of patients.

Table 6

<table>
<thead>
<tr>
<th>Survival of Patients with Type III Dissection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients who were not operated</td>
</tr>
<tr>
<td>Died during initial episode</td>
</tr>
<tr>
<td>Discharged alive after initial episode</td>
</tr>
<tr>
<td>Subsequently died (11 mo to 5 yr)</td>
</tr>
<tr>
<td>Alive (2 and 3 yr)</td>
</tr>
<tr>
<td>Patients who were operated</td>
</tr>
<tr>
<td>Died postoperatively (0 to 17 days)</td>
</tr>
<tr>
<td>Alive (6 mo to 4 yr)</td>
</tr>
<tr>
<td>Aortic dissection, an incidental autopsy finding</td>
</tr>
<tr>
<td>Total</td>
</tr>
</tbody>
</table>

Discussion

Present information regarding the course of patients with aortic dissection is gleaned from retrospective analysis of autopsied cases,\(^1, 13, 15-19\) of surgical cases,\(^2\) of patients seen in referral centers,\(^2, 7, 8, 10, 11, 16\) or of cases collected from the literature.\(^1, 13\) Bias is inherent in such data. Autopsy material is weighted in favor of the more severely ill patients. Surgical series do not include pa-
patients who die before they can be operated on, and many patients succumb before reaching a referral center. Unusual cases or cases with a successful outcome more commonly find their way into the literature and may bias collected series.

The outlook may be less grim than is presently believed since the diagnosis is often unsuspected or unproven in patients with less extensive or painless dissection. Cases discovered incidentally at autopsy support the idea that prolonged survival following unrecognized disease occasionally occurs.

The present study does not escape all of the biases mentioned. Prior to 1960, the diagnosis was acceptably proved short of autopsy in only a few instances. All the objections to the analysis of a series of autopsy cases apply. Subsequently, the diagnosis has been confirmed during life in 12 instances by aortography. Thus the group of patients collected since 1960 may be more representative. Our collection of patients does have the virtue of being the total experience of a large general hospital. Most patients were seen within a few hours of the onset of symptoms. The inherent selectivity in the clinical material of referral centers is thus avoided.

Several features of the natural history of dissecting aortic aneurysm which have not been previously emphasized are highlighted by the present review. The long-term outlook for patients whose aortic dissection was limited to the descending aorta was better in our study group than has been reported by others.\textsuperscript{7, 8, 10, 14} This is true in those instances in which surgery was not done. Six of 12 such patients who were not operated on survived to leave the hospital. Survival for 11, 24, 26, 34, 41, and 69 months is known in those six patients. Death in three was unrelated to aortic dissection and in a fourth was attributable to an attempt to replace the descending aorta because of the chronic dissection 69 months after the acute episode. Two are alive more than 2 years after their acute illness.

The better early survival rate of patients whose aneurysms do not involve the ascending aorta has been noted by others,\textsuperscript{1, 3, 7, 13--20} but the effect of this fact on the composition of series of surgically treated cases has not been emphasized. Although the ascending aorta is involved in the majority of patients who are seen initially (65\% of the present series), selective attrition early in the disease precludes operative intervention in many of this group. In our own experience, only seven of the 22 survivors at the end of 1 week after onset had evidence of involvement of the ascending aorta.

The available reports describing the results of surgical treatment indeed show improved survival when operated cases are compared with series of autopsied cases.\textsuperscript{2, 8, 21} This is not surprising when it is considered that patients with dissection limited to the descending aorta make up 55 to 80\% of the surgical groups cited. Thus surgical series often consist of individuals more likely to survive even without operation and are in no way comparable to series of autopsied cases.

The extent of dissection is not reported in the descriptions of the vigorous approach to therapy with antihypertensive agents.\textsuperscript{11, 12} Aortic dissection limited to the descending aorta may be more common than was previously supposed. The usual clues suggesting dissecting hematoma as the etiology of a patient’s pain were frequently missing in this group. A pulsatile sternoclavicular joint\textsuperscript{22} was often the only sign of the true nature of the

\begin{table}
\centering
\caption{Cause of Death in Type III Dissecting Aortic Aneurysm}
\begin{tabular}{ll}
\hline
Cause of death & No. of patients \\
\hline
External rupture & 5 \\
Left hemothorax & 5 \\
Hemomediastinum & 1 \\
Postoperative complications & 4 \\
Cerebrovascular accident & 3 \\
Pneumonia & 1 \\
Hypertensive encephalopathy & 1 \\
Cardiac arrest at surgery & 1 \\
Alive & 4 \\
Total & 19 \\
\hline
\end{tabular}
\end{table}
Dissecting Aneurysm of the Aorta

Dissecting aneurysm of the aorta appears to be far more likely to survive undiagnosed.

Contrast radiography now allows the definitive diagnosis of this type of aneurysm during life. Prior to 1960, 31 instances of aortic dissection were found to be suitable for the present study. Only six type III dissections (19%) were included in this essentially autopsy series but 13 of 31 cases (42%) evaluated thereafter were of this variety. Retrograde aortography was first used in this institution for this purpose in 1960. It has been diagnostic in 10 instances of type III and in two instances in type I dissection. We believe that increased awareness of this disease on the part of our staff and the introduction of contrast radiography reduced the number of type III dissections which are unsuspected or unproved.

The division of patients with dissecting aortic aneurysm into diagnostic and therapeutic groups with regard to the extent of the disease process appears justified. It seems established that when the ascending aorta is involved, as it is in a majority of cases, the disorder is likely to be fatal within a few days usually from rupture into the pericardial space. Aortic regurgitation, major arterial occlusion, and signs of neurological illness are often present. If the disease is considered, the diagnosis is usually obvious. Heroic, lifesaving measures are justified.

We would suggest that when the medial dissection begins distal to the arch vessels, the second most common point of origin, the classic physical findings are usually absent and the diagnosis is frequently not considered in the early stages. These patients commonly, and perhaps usually, survive for weeks or months. A quarter or more of untreated patients may survive for months or years to die of other causes.

Despite the current study we feel that the natural history of this disorder is not completely understood and that the available data do not allow definitive conclusions regarding the efficacy of any form of therapy. There is a clear need for carefully controlled studies of the therapy of this problem which take into account the extent of the dissecting process.

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12. Editor's Note: Following Pomerantz, M., Young, W. G. Jr., and Seely, W. C. (See reference 9.)
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Circulation. 1967;35:880-888
doi: 10.1161/01.CIR.35.5.880
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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