Behavior of mortality due to aortic dissection in Cuba

Osvaldo Valdés Dupeyrón\textsuperscript{a,b}, MD, MSc; José Hurtado de Mendoza Amat\textsuperscript{c}, PhD; Teresita de J. Montero González\textsuperscript{c}, PhD; Reynaldo Álvarez Santana\textsuperscript{d}, MD; Antonio de Arazoza Hernández\textsuperscript{a}, MD, MSc; and Jean L. Chao García\textsuperscript{a}, MD

\textsuperscript{a} Cardiology Hospital of the Center for Medical and Surgical Research (CIMEQ). Havana, Cuba.
\textsuperscript{b} Dr. Luis Díaz Soto Hospital. Havana, Cuba.
\textsuperscript{c} Department of Anatomical Pathology. Dr. Luis Díaz Soto Hospital. Havana, Cuba.
\textsuperscript{d} Department of Anatomical Pathology. Hermanos Ameijeiras Hospital. Havana, Cuba.

ABSTRACT

Introduction: Aortic dissection is a disease of poor prognosis, with a high mortality rate, even when it is diagnosed early and the adequate surgery is performed.

Objectives: To describe some characteristics of the patients who died due to aortic dissection in Cuba.

Method: A retrospective cross-sectional study was conducted. It included 888 deceased patients with a diagnosis of aortic dissection, who were registered in the Automated Registration and Control System of Anatomical Pathology (SARCAP, for its acronym in Spanish) in Cuba, from 1962 to 2004.

Results: The age groups from 65 to 74 and from 75 to 84 years predominated, with 266 (30.0%) and 210 (23.6%) deaths, respectively. The largest number of deaths [496 (55.9%)] occurred in the first 24 hours. Stanford type A aortic dissection was the most frequent type [535 diagnoses were made (61.1%)]. The most common underlying cause of death was aortic dissection itself (61.6 %) and the most common direct cause of death was hemopericardium (43.9%). The rate of diagnostic agreement was 33.5 % in the underlying cause and 28.8 % in direct cause.

Conclusions: During the 42 years covered by the study, the patients who died from aortic dissection in Cuba predominantly had Stanford type A dissections, were over 55 years of age and had a hospital stay of less than 2 months. The largest number of deaths occurred in the first 24 hours and the rate of diagnostic agreement was low.

Key words: Aortic dissection, Necropsy, Diagnostic agreement, SARCAP

Comportamiento de la mortalidad por disección aórtica en Cuba

RESUMEN

Introducción: La disección aórtica es una enfermedad de pronóstico muy reservado, con una elevada mortalidad, aun cuando se diagnostique precozmente y se realice la intervención quirúrgica adecuada.

Objetivo: Describir algunas características de los fallecidos por disección aórtica en Cuba.
Valdés Dupeyrón O, et al.

**Método:** Se realizó un estudio retrospectivo de corte transversal, que incluyó 888 fallecidos con diagnóstico de disección aórtica, incluidos en el Sistema Automatizado de Registro y Control de Anatomía Patológica (SARCAP) en Cuba, desde el año 1962 hasta el 2004.

**Resultados:** Predominaron los grupos etarios de 65-74 y 75-84 años, con 266 (30,0 %) y 210 (23,6 %) defunciones, respectivamente. El mayor número de muertes [496 (55,9 %)] ocurrió en las primeras 24 horas. La disección aórtica tipo A de Stanford fue la de mayor frecuencia [535 diagnósticos (61,1 %)]. La causa básica de muerte más encontrada fue la propia disección aórtica (61,6 %) y la causa directa, el hemopericárdico (43,9 %). El índice de coincidencia diagnóstica fue de 33,5 % en la causa básica y de 28,8 % en la directa.

**Conclusiones:** Los fallecidos por disección aórtica en Cuba, durante los 42 años estudiados, tuvieron predominantemente una disección tipo A de Stanford, más de 55 años de edad y una estadía hospitalaria menor de 2 meses. El mayor número de muertes ocurrió en las primeras 24 horas y el índice de coincidencia diagnóstica fue bajo.

**Palabras clave:** Disección aórtica, Necropsia, Coincidence diagnóstica, SARCAP

**INTRODUCTION**

Aortic dissection (AD) was described by the renowned Italian anatomist and pathologist Geovani Batista Morgagni in 1761. It was studied and defined as dissecting aneurysm by the French physician René Théophile Hyacinthe Laenec in 1819, and was first diagnosed clinically by Swaine and Latham in 1885 and 1886, respectively. It is a tear in the intima of the vessel, exposing the previously ill tunica media to intraluminal systolic blood force. Blood enters the media layer and splits the vessel wall into two layers. The blood-filled space in the dissected layers becomes the false lumen. This dissection extends along a variable length in the aorta, usually in a forward direction, and, at times, in a retrograde direction, from the site of intimal tear.

The term dissecting aneurism is still used by some medical professionals nowadays; however, this concept has been replaced with the term aortic dissection for decades. The aneurysm and AD are independent diseases. The latter may generally occur in an acute form, although, after the tear, there is a subsequent gradual expansion of the weakened outer wall of the aorta. On the other hand, the gradual deterioration of the aortic wall in aneurysms may cause a chronic dissection. Therefore, the term dissecting aneurysm should be reserved only for this possibility.

The concept of acute aortic syndrome was introduced late last century. It is defined as an acute process in the aortic wall that determines a high risk of rupture with a high morbidity and mortality. It includes AD, intramural hematoma, penetrating ulcer, and other diseases such as symptomatic or ruptured thoracic aneurysm, aortic transection and aorto-pulmonary fistula, as an erosive complication of the aneurism.

Currently, AD is considered a catastrophic illness, with an estimated incidence of 5 to 30 cases per million inhabitants. About 10 000 patients a year suffer from aortic dissection in the United States. Without surgical treatment, its natural progression is fatal. A significant number of patients die without hospital medical attention. AD diagnosis is not made in 38% of patients and it reaches 28% in necropsies. Death by AD may be related to aortic rupture, cardiac tamponade, severe acute aortic regurgitation, or acute myocardial infarction by coronary involvement. The presence of recurrent pain, shock, cardiac arrest, and a pericardiocentesis with bloody fluid indicate the possible existence of an AD of the ascending aorta breaking into pericardial cavity.

Several studies on AD have been conducted in our country, including clinical trials and post-mortem studies, which have shown the incidence and prevalence of the disease, as well as the fundamental aspects of its diagnosis and treatment. For this reason, it was decided to conduct this study, with the aim of covering an almost complete sample of our country, in more than four decades, thus providing continuity to previous research on this disease.

**METHOD**

A retrospective cross-sectional study was conducted. It included 888 autopsy reports with a diagnosis of AD from the Automated Registration and Control System of Anatomical Pathology (SARCAP, for its acronym in...

The AD was classified according to the Stanford University anatomical and pathophysiological classification, proposed by Daily et al in 1970, which includes:
- Type A: dissections involving the ascending aorta, which correspond to DeBakey types I and II.
- Type B: dissections that do not affect the ascending aorta, which includes DeBakey type III.

The following criteria were used for clinicopathological correlation:
- Yes: When the correlation was total or partial.
- No: When there was no correlation or data were insufficient.

Univariate and bivariate frequency distributions were built, with graphical representation. Chi-square homogeneity tests and proportion test for paired samples, with mutually exclusive characteristics, were used. Hypothesis tests and estimation required random samples. If the randomness of the sample cannot be justified, then the results of the tests are unreliable; however, their information may be used to help decide with what is observed in Tables 4 and 5.

RESULTS

Our study included all autopsies performed in patients over 15 years of age in 43 hospitals across the country, from 1962 to 2004, with a total of 101,082 autopsies, performed by dozens of specialists in Anatomical Pathology. In the first three years no cases of AD were reported, although it should be noted that the first five years included the deaths from a single hospital. Subsequently, other hospitals were incorporated to form the current network. The years with the highest incidence of this disease were 1994 with 133 and 2000 with 103 autopsies.

According to the type of AD, following the Stanford classification (Table 1), there was a predominance of type A, with 543 deaths (61.1%), of which 253 (28.5% of the total) were classified as DeBakey type I and 290 as type II (32.6% of the total). In 390 autopsies, pericardial effusion was found, due to the rupture of the dissection into that cavity. Stanford type B and DeBakey type III dissections were found in 345 deceased patients (38.9%), including 61 ruptures into pleura and 81 into the peritoneal cavity.

The mean age was 69 years, ranging between 20 and 100 years (Table 2). Moreover, there is a relationship between AD and increasing age, with a higher number of cases over 50 years of age. The most affected age groups were those from 65-74 and 75-84 years, with 266 (30.0%) and 210 (23.6%) deaths, respectively.

With regard to hospital stay (Table 3), the largest number of deaths occurred during the first 24 hours, with a total of 496 deceased patients (55.86%), follow-

<table>
<thead>
<tr>
<th>Age groups (years)</th>
<th>Nº</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>15-24</td>
<td>1</td>
<td>0,1</td>
</tr>
<tr>
<td>25-34</td>
<td>3</td>
<td>0,3</td>
</tr>
<tr>
<td>35-44</td>
<td>26</td>
<td>2,9</td>
</tr>
<tr>
<td>45-54</td>
<td>76</td>
<td>8,6</td>
</tr>
<tr>
<td>55-64</td>
<td>177</td>
<td>19,9</td>
</tr>
<tr>
<td>65-74</td>
<td>266</td>
<td>30,0</td>
</tr>
<tr>
<td>75-84</td>
<td>210</td>
<td>23,6</td>
</tr>
<tr>
<td>85-94</td>
<td>108</td>
<td>12,2</td>
</tr>
<tr>
<td>≥ 95</td>
<td>5</td>
<td>0,6</td>
</tr>
<tr>
<td>Non-specified age</td>
<td>16</td>
<td>1,8</td>
</tr>
<tr>
<td>Total</td>
<td>888</td>
<td>100</td>
</tr>
</tbody>
</table>

Source: SARCAP

Table 1. Distribution of the sample according to Stanford and DeBakey classifications.

<table>
<thead>
<tr>
<th>Stanford Type A</th>
<th>Stanford Type B</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nº</td>
<td>%</td>
<td>Nº</td>
</tr>
<tr>
<td>DeBakey I</td>
<td>253</td>
<td>28,5</td>
</tr>
<tr>
<td>DeBakey II</td>
<td>290</td>
<td>32,6</td>
</tr>
<tr>
<td>Total</td>
<td>543</td>
<td>61,1</td>
</tr>
</tbody>
</table>

Source: SARCAP

Table 2. Sample distribution by age groups.

Table 3. Sample distribution by age groups.
ed by the deaths occurred between the first and third day of hospital stay, with 209 cases (23.86 %), making a cumulative of 79.39 %. This parameter increased with the course of days to reach 99.9 % at 60 days, that is, 99.9 % of patients died within two months. One patient had a longer hospital stay. Mortality increased in the course of time.

Table 4 shows the distribution of the sample according to underlying and direct causes of death. Regarding the underlying cause, AD had the highest incidence. It was found in 541 death certificates (61.6 %), followed by atherosclerosis (14.3 %), cystic medial degeneration (13.4 %) and finally hypertension (9.2 %). That is, 98.5 % of the underlying causes of death were vascular alterations, which are closely related to the genesis of aortic syndromes.

With regard to the direct cause of death, hemopericardium was the most common one, found in 390 deaths (43.9 %); followed by shock, which was diagnosed in 164 autopsies (18.5 %). Pleural effusion was the third with 38 deaths (4.3 %), and finally bronchopneumonia and acute anemia, found in 37 autopsies (4.2 % each one).

Regarding the diagnostic agreement on the direct cause of death (Table 5), there was full and partial clinicopathological correlation only in 33.5 % of the sample. It was similar with the underlying cause of death, with a correlation of 28.8 %. These results show high rates of diagnostic discrepancy.

**DISCUSSION**

With the decline in autopsies during the last three decades in most countries of the world, obtaining data from a large number of cases is very complicated. Despite the medical-legal and educational importance of this procedure, its use is becoming increasingly scarce18-20. In Cuba, it has been possible to maintain acceptable levels in postmortem studies, although we are not exempt from this crisis. From 1991 to 2011, the rate of autopsies in our country, not including fetuses, was greater than 33 %; and, in the case of hospital deaths, it has remained near 60 % over the same period21.

At present, there are numerous classification systems to describe AD, although Stanford classification main-
tains hegemony due to its usefulness from a physio-pathological point of view. The involvement of the ascending aorta influences the prognosis and the subsequent course of action. That is, when the tearing includes the ascending aorta, surgery is always required. Conversely, if the dissection does not affect this segment, the course of action may be medical or endovascular treatment.

The AD is a disease that mainly affects elderly people. Our results are similar to those found in studies such as the prestigious IRAD and RESA, where there is a prevalence of patients over 60 years, probably due to the loss of aortic elasticity and distensibility associated with aging. Histologically, senile aorta shows a fragmentation of elastin and a concomitant increase in collagen, which favors a physiological reduction of distensibility and increases myocardial oxygen consumption by 20 to 40%. Recent experimental data from animal models suggest that impaired vasa vasorum flow to the aortic wall leads to increased rigidity with histological changes similar to ageing.

Without surgical treatment, AD mortality increases by 1% every hour after the onset of the symptoms. As a result, more than one third of patients with this disease die within the first 24 hours, half of them in the first 48 hours, two thirds in the first two weeks and nearly 90% in the first 3 months. The risk of death is increased in patients with complications such as aortic rupture, stroke, visceral ischemia, cardiac tamponade, and circulatory failure. Data from other important registries on acute AD show that in the absence of immediate surgical repair, medical treatment is associated with high mortality. Even with surgical repair, the hospital mortality rates are 10% after the first day, 12% at 2 days and almost 20% at 2 weeks.

In our study, in about 40% of deaths, AD was not identified as the underlying cause of death. The cystic and atherosclerotic degeneration of the media and hypertension are included in this group. Cystic medial degeneration is defined as a degenerative process of the tunica media, with loss or fragmentation of elastic fibers and smooth muscle cells. Most dissections in young people are generally due to congenital abnormalities of the connective tissue, affecting the media of the aorta. The atherosclerotic degeneration is the alteration of the intima, with formation of large plaques that end up destroying the elastic fibers and smooth muscle cells of the media, which in turn causes weakness and dilation of the arterial wall. Hypertensive disease increases the absolute stress on the arterial wall, the force of left ventricular ejection (dP/dt) and myocardial oxygen consumption. All this intensifies chronic damage of the arterial wall and the risk of dissection or rupture.

In this study, the increased incidence of hemopericardium is related to the predominance of Stanford type A dissections. The accumulation of blood in the pericardial sac has an anatomical relationship with the acute aortic syndrome, because the majority of dissections affect the ascending aorta. In 65% of cases the site of intimal tear is located there, mostly within the first 5 inches. The tear may progress and break into the pericardial cavity, which increases the volume and pressure within it. These alterations, when they occur in an acute form, cause the pericardium to tense up and lose its elasticity. When the intrapericardial pressure exceeds intracavitary pressure, circulatory collapse occurs and death is inevitable if an emergency evacuation of fluids is not performed. About 10% of patients with a diagnosis of Stanford type A dissection suffer from cardiac tamponade with a high risk of death.

On the other hand, the shock is linked to blood loss, pain and heart failure is secondary to acute aortic regurgitation or acute myocardial infarction. Pleural effusion is related to the dissection of the thoracic aorta and is more likely to occur in the left pleura. Bronchopneumonia was found in patients who survived the early days, and is due to septic in-hospital complications, because is not directly related to the cause of the admission to hospital, in intensive care units most of the times.

The AD has well-defined clinical symptoms, but its acute onset may mislead the physician. In most cases, there is a diagnosis of myocardial infarction, pulmonary embolism, pericarditis and other conditions. The clinicopathological discrepancies range between 25 and 52%, but when the aortic syndromes are analyzed, these percentages are close or even above 50%. found a discrepancy rate of 58.8% in the aortic aneurysm, Cameron and McGoogan, 57.9% and Fares et al., 64.2%; these results are in agreement with our study. Furthermore, in a study conducted in the province of Cienfuegos, on 55 deaths due to AD, the diagnosis made when the patient was admitted to hospital was confirmed in only 18.18% of cases, however, Battle et al. found a discrepancy.
rate of 28.1%.

CONCLUSIONS
During the 42 years covered by the study, those who died from AD in Cuba predominantly had Stanford type A dissections, were over 55 years of age and had a hospital stay of less than 2 months. The largest number of deaths occurred in the first 24 hours and the rate of diagnostic agreement was low.

REFERENCES
22. Ladouceur M, Fermanian C, Lupoglazoff JM, Edou-
23. Contreras Zúñiga E, Zuluaga Martínez SX, Gómez Mesa JE, Ocampo Duque V, Urrea Zapata CA. Disec-
ción aórtica: estado actual. Rev Costarric Cardiol.
25. Ford PF, Farber MA. Role of endovascular therapies in the management of diverse thoracic aortic pathology. Perspect Vasc Surg Endovasc Ther. 2007;
14-8.
II312-17.
32. Senay S, Alhan C, Toraman F, Karabulut H, Dagde-
len S, Cagil H, et al. Endovascular stent-graft treat-
457-60.
33. Ince H, Nienaber CA. Diagnosis and management of patients with aortic dissection. Heart 2007;93(2):
266-70.
34. Nienaber CA, Eagle KA. Aortic dissection: new frontiers in diagnosis and management. Part II: Therape-
35. Santini F, Luciani GB, Montalbano G, Messina A,
Faggian G, Mazzucco A. Acute type A aortic dis-
ASA/SCA/SCAI/SIR/STS/SVM guidelines for the di-
agnosis and management of patients with Thora-
cic Aortic Disease: A report of the American College of Cardiology Foundation/American Heart Associa-
38. Shojania KG, Burton EC, McDonald KM, Goldman L. Changes in rates of autopsy-detected diagnostic errors over time: A systematic review. JAMA. 2003;
289(21):2849-56.
40. Spiliopoulos C, Papadodima S, Kotakidis N, Kuotsel-