Aneurysmal Coronary Artery Disease

PAUL S. SWAYE, M.D., LLOYD D. FISHER, PH.D., PAUL LITWIN, B.A.,
PAUL A. VIGNOLA, M.D., MELVIN P. JUDKINS, M.D., HARVEY G. KEMP, M.D.,
J. GERARD MUDD, M.D., AND ARTHUR J. GOSSELIN, M.D.

SUMMARY To examine the clinical and historical features and the natural history of aneurysmal coronary disease, we reviewed the registry data of the Coronary Artery Surgery Study (CASS). Nine hundred seventy-eight patients, representing 4.9% of the total registry population, were identified as having aneurysmal disease. No significant differences were noted between aneurysmal and nonaneurysmal coronary disease patients when features such as hypertension, diabetes, lipid abnormalities, family history, cigarette consumption, incidence of documented myocardial infarction, presence and severity of angina, and presence of peripheral vascular disease were examined. In addition, no difference in 5-year medical survival was noted between these two groups. These findings suggest that aneurysmal coronary disease does not represent a distinct clinical entity but is, rather, a variant of coronary atherosclerosis.

ANEURYSMAL coronary disease is characterized by abnormal dilatation of a localized or diffuse segment of the coronary arterial tree. This involvement has also been termed coronary ectasia or dilating arteriosclerosis; destruction of the vessel media is the usual histologic feature. Frequently, aneurysmal disease coexists with coronary atherosclerosis and has raised the question of whether aneurysmal disease is a variant of atherosclerotic coronary disease or a distinct entity. The presence of dilated coronary segments, even in the absence of obstructive disease, is believed to result in alterations in blood flow and stasis, which predispose these patients to myocardial ischemia and infarction.

Although many reports on aneurysmal coronary disease have been published, the number of patients in each report has been small.1-14 We therefore evaluated a large group of patients with aneurysmal coronary disease from the registry data of the multinstitutional Coronary Artery Surgery Study (CASS), sponsored by the National Institutes of Health (NIH).15 We examined the clinical and historical features and the natural history of coronary aneurysmal disease to determine whether it represents a distinct clinical entity or a variant of coronary atherosclerotic disease.

Materials and Methods

Between July 1975 and May 1979, clinical, laboratory and angiographic data were collected in a standardized fashion and entered into a registry of consecutive patients undergoing coronary arteriography for clinically suspected coronary artery disease at 15 participating clinical centers of CASS. Patients who were subsequently shown to have normal coronary arteries were included in the registry. Patients studied for suspected coronary artery disease who were found to have another form of heart disease were excluded. Those who underwent coronary angiography for evaluation of other conditions, such as valvular heart disease, cardiomyopathies and congenital heart disease, were also excluded even if subsequent evidence showed that coronary artery disease was a major clinical problem.

A total of 20,087 patients from the participating institutions was entered into the CASS registry. The patients who had undergone cardiac surgery were not included. Each patient was interviewed at the time of hospitalization for coronary angiography by a trained data technician or the responsible physician. The base-


From the Miami Heart Institute, Miami Beach, Florida; Loma Linda University, Loma Linda, California; St. Luke’s Hospital, New York, New York; St. Louis University, St. Louis, Missouri; and the Coordinating Center for Collaborative Studies in Coronary Artery Surgery, University of Washington, Seattle, Washington.

Address for correspondence: Paul S. Swayne, M.D., Miami Heart Institute, 4701 North Meridian Avenue, Miami Beach, Florida 33140.
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line historical, physical and laboratory data and the results of coronary arteriography and left ventriculography were collected and recorded on appropriate data forms.

**Clinical Data Base**

A number of clinical descriptors were obtained when the patients entered the registry. The predominant symptoms included chest pain, dyspnea, orthopnea, paroxysmal nocturnal dyspnea, palpitations and fatigue. Chest pain was classified by the responsible physician as definitely angina, probably angina, probably not angina or definitely not angina. Functional classification was designated according to the Canadian Cardiovascular Society. Family history was considered positive when there was knowledge of symptoms of angina pectoris or history of myocardial infarction in parents, siblings or aunts or uncles related by blood before age 55 years. A history of cigarette smoking, treated hypertension, diabetes, lipoprotein abnormalities, or documented myocardial infarction was recorded.

The arteriographic data forms allowed entry of information on 27 segments of the coronary vasculature bed. The participating sites agreed to use a uniform code to describe coronary artery anatomy. In this way, angiographic data from different clinical sites could be compared. Details of the classifications used have already been reported. Briefly, coronary stenoses were read as the percent narrowing of the maximal luminal diameter for each segment. The criteria for clinically significant coronary artery obstruction were defined as either a 70% or more reduction in the internal diameter of the right, left anterior descending or left circumflex coronary artery or 50% or greater reduction in the internal diameter of the left main coronary artery. The presence of coronary aneurysmal dilatation was recorded. Discrete or diffuse aneurysmal disease was defined as coronary dilatation that exceeded the diameter of normal adjacent segments or the diameter of the patient’s largest coronary vessel by 1½ times (fig. 1).

Left ventricular function was assessed from ejection fractions calculated by a single-plane adaptation of the area-length method of Sandler and Dodge. In addition, a left ventricular wall motion score was obtained by analyzing the five left ventricular segments visualized on right anterior oblique ventriculograms.

Uniformity of interpretation of angiographic data among the 15 contributing institutions was achieved by designating quality control sites as part of the ongoing CASS project. Each month, the Coordinating Center randomly selected from each clinical site three films depicting coronary lesions. These films were sent to a quality control site to be reinterpreted. The readings from the original site and quality control site were compared and the discrepancies recorded. By the end of enrollment, 871 films were read as part of this study. As part of the present study, three of the authors independently reviewed 54 films, selected at random from the clinical sites, designated as demonstrating aneurysmal disease. They reviewed these films to evaluate the reproducibility of criteria for aneurysmal disease. The result of this analysis revealed complete agreement in 70.2% of the randomly selected films. In 20.4% there was disagreement with the original interpretation and in 9.4% the findings were equivocal.

**Results**

Of the 20,087 patients, 978 (4.9%) were identified as having aneurysmal coronary dilatation (fig. 2). Twenty-one had aneurysmal disease and no coronary stenosis (group A), 69 had aneurysmal disease and less
than 70% stenosis (group B), and 888 had aneurysmal disease and 70% stenosis or greater (group C). The control population in this study consisted of the 19,109 other patients (95.1%) in the CASS registry.

Among the 978 patients in the study group, the proximal and middle segments of the right coronary artery were most frequently involved by aneurysmal disease. The next most frequently involved segments were the proximal left anterior descending and circumflex coronary arteries.

The 978 patients with aneurysmal disease were compared with a group of control patients without aneurysmal disease who had any degree of coronary stenoses (n = 15,249). Patients with completely normal coronary arteries were excluded from this comparison group.

The percentage of males with aneurysmal disease was significantly greater than that in the control group (88.2% vs 80.1%, p < 0.001). No age differences were noted. No differences were noted with regard to coronary disease risk factors such as hypertension, diabetes, lipid abnormalities and cigarette use. However, a family history of coronary disease was more common in the control group (43.2% vs 38.5%, p < 0.05). Patients with aneurysmal disease had a higher incidence of documented myocardial infarction (52.9% vs 47.0%, p < 0.001). However, no differences were noted with regard to other clinical descriptors such as angina, peripheral vascular disease and Canadian Heart Association functional classification. The incidence of three-vessel coronary disease was higher in the aneurysmal group (42.3% vs 34.2%, p < 0.001); however, left ventricular function, as measured by ejection fraction and left ventricular wall motion score, was similar in the two groups.

Patients with aneurysmal disease and no associated coronary stenosis or nonsignificant coronary stenosis (groups A and B, n = 90) were then compared with a population that did not have aneurysmal disease or significant coronary stenosis (n = 1893). Patients with normal coronary arteries were excluded from this comparison group. Although the percentage of males in the aneurysmal group was significantly greater than in the controls (75.3% vs 62.2%, p < 0.05), no differences were noted with regard to age, family history of coronary disease, hypertension, diabetes, lipid abnormalities, and cigarette smoking, associated angina, prior myocardial infarction, functional classification, incidence of peripheral vascular disease, left ventricular ejection fraction and left ventricular wall motion score.

Cumulative survival curves* demonstrated reduced survival at the 5-year medical follow-up in the patients with aneurysmal disease and any degree of coronary stenosis (groups B and C) compared with patients without aneurysmal disease and any degree of coronary obstruction (74% vs 83%) (fig. 3). Further analysis of these groups was performed taking into account arteriographic and ventriculographic findings, including the combinations of number of vessels diseased, the number of proximal vessels diseased and the LV score. The survival curve (fig. 4) failed to show a significant difference between the two groups. Similarly, the survival curves of medically treated patients with aneurysmal disease and either no or nonsignificant coronary stenosis (groups A and B) were not different from those in a control group without aneurysmal disease or significant coronary stenosis (fig. 5).

*Computed by the life-table or actuarial method; the statistical significance of differences between groups was compared by log-rank test.
The patients with aneurysmal disease who underwent coronary bypass surgery (n = 496) were then compared with a population of patients without aneurysmal disease (n = 7622) who also underwent bypass surgery. The 5-year cumulative survival curves showed no significant differences between the two groups.

**Discussion**

Aneurysmal dilatation of the coronary arteries was first described by Bourgon in 1812. In 1929, Packard and Wechsler reviewed 21 previously reported cases. Since then, numerous isolated case reports and small series of patients have been described. The CASS registry, sponsored by the NIH, affords a unique opportunity to study a large group of patients with angiographically documented coronary aneurysmal dilatation so that the clinical and historical features as well as the natural history of this disease can be further elucidated.

In the current study, patients with aneurysmal coronary disease had a similar incidence of "coronary" risk factors such as hypertension, diabetes, lipid abnormalities and cigarette use compared with controls. This finding differs from other studies in which a higher incidence of systemic hypertension was noted in patients with aneurysmal disease. Markis et al. speculate that the systemic hypertension might play a role in the pathogenesis of this disease entity. The reason for this difference is not clear but may relate to differences in patient selection. Other investigators also failed to demonstrate a higher incidence of "coronary" risk factors, including hypertension, in patients with aneurysmal coronary disease.

In our study, patients with aneurysmal disease had a greater incidence of documented myocardial infarction than the control group, as observed by others. However, our patients with aneurysmal disease and nonsignificant coexistent obstructive lesions did not have a greater incidence of infarction. It would be tempting to suggest that an aneurysmal segment with its possible altered flow characteristics in association with a significant obstruction predisposes to myocardial infarction. However, the patients with aneurysmal disease...
and significant coronary stenoses, who made up the overwhelming majority of the aneurysm group, had a significantly higher incidence of three-vessel involvement. Therefore, the increased incidence of infarction might merely reflect the increased incidence of three-vessel disease in this aneurysmal group. Furthermore, when other factors that affect survival, such as percent luminal narrowing and left ventricular wall motion score, were taken into account, no real differences in survival existed between the patients with aneurysmal disease and comparable patients without aneurysmal disease.

In conclusion, our findings failed to reveal a clinical picture or pattern of risk factors that would suggest a diagnosis of aneurysmal coronary disease. Aneurysmal coronary disease appears to be a variant of occlusive coronary atherosclerosis, and both lesions frequently coexist in the same vessel.

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References


Appendix

Cooperating Clinical Sites


Central Electrocardiographic Laboratory

University of Alabama: L. Thomas Sheffield, M.D.,* David Roitman, M.D., and Carol Troxell, B.S.

Coordinating Center

University of Washington: Lloyd Fisher, Ph.D.,* Mary Jo Gillespie, M.S., Kathryn Davis, Ph.D., J. Ward Kennedy, M.D., and Richard Kronmal, Ph.D.

Chairman of Steering Committee

Thomas Killip, M.D., Henry Ford Hospital.

*Denotes principal investigator.
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P S Swayne, L D Fisher, P Litwin, P A Vignola, M P Judkins, H G Kemp, J G Mudd and A J Gosselin

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