JOURNAL OF THE AMERICAN COLLEGE OF CARDIOLOGY
© 2015 BY THE AMERICAN COLLEGE OF CARDIOLOGY FOUNDATION
PUBLISHED BY ELSEVIER INC.

VOL. 65, NO. 22, 2015
ISSN 0735-1097/$36.00
http://dx.doi.org/10.1016/j.jacc.2015.03.569

Prognostic and Bioepidemiologic Implications of Papillary Fibroelastomas

Syahidah S. Tamin, MBBS,* Joseph J. Maleszewski, MD,*† Christopher G. Scott, MS,‡ S.K. Khan, MBBS,§ William D. Edwards, MD,*† Charles J. Bruce, MD,* Jae K. Oh, MD,* Patricia A. Pellikka, MD,* Kyle W. Klarich, MD*

ABSTRACT

BACKGROUND Papillary fibroelastomas (PFE) are benign neoplasms with little available outcome data.

OBJECTIVES This study sought to describe the frequency and clinical course of patients with surgically removed PFE and echocardiographically suspected, but unoperated, PFE.

METHODS Mayo Clinic pathology and echocardiography databases (January 1, 1995, to December 31, 2010) were queried, resulting in 511 patients: group 1 (n = 185), including patients with surgically removed, histopathologically confirmed PFE; group 1a (n = 94; 51%) with PFE removed at primary surgery; and group 1b (n = 91; 49%) with PFE removal at time of another cardiac surgery. Group 2 (n = 326) patients had echocardiographic evidence of PFE but no cardiac surgery to remove PFE.

RESULTS Group 1 had mean age of 63 ± 14 years (116 women [63%]). During the study period, we identified 112 cardiac myxomas in the pathology database and 142 in the echocardiographic database. Mean age in group 2 was 67 ± 14 years (162 women [50%]). PFE occurred most commonly on cardiac valves (n = 400 [78%]). In group 1, transient ischemic attack or stroke was the presenting symptom in 58 patients (32%). With surgical removal of valvular PFE, the valve was preserved in 92 (98%). Recurrence was documented in 3 patients (1.6%). Follow-up stroke risk in groups 1, 1a, and 1b at 1 year was 2%, 0%, and 4%; at 5 years, 8%, 5%, and 11%, respectively. Cerebrovascular accident risk in group 2 at 1 and 5 years was 6% and 13%.

CONCLUSIONS In patients with echocardiographically suspected PFE who do not undergo surgical removal, rates of cerebrovascular accident and mortality are increased. (J Am Coll Cardiol 2015;65:2420–9) © 2015 by the American College of Cardiology Foundation.

Cardiac papillary fibroelastomas (PFE) are small, benign endocardial lesions that are clinically important because of their documented embolic potential. Although generally considered a common benign cardiac tumor, they are not the most common. In 1 autopsy series (1), PFE were the third most frequently occurring cardiac tumor, behind cardiac myxoma and lipoma. Histologically, PFE are avascular structures composed of fibroelastic tissue surrounded by endocardium, imparting a characteristic echocardiographic appearance (2–4).

Historically, PFE were diagnosed incidentally at autopsy, thus reflecting probable underdiagnosis. With increasing use of echocardiography, improved resolution, and transesophageal techniques, more PFE are now being detected antemortem. We sought to better elucidate the true frequency of PFE and to define clinical characteristics and outcomes of surgically treated, histopathologically confirmed cases in a single, large clinical practice. Additionally, we sought to better understand the clinical course of PFE suspected on

From the *Division of Cardiovascular Diseases, Mayo Clinic, Rochester, Minnesota; †Division of Anatomic Pathology, Mayo Clinic, Rochester, Minnesota; ‡Division of Biomedical Statistics and Informatics, Mayo Clinic, Rochester, Minnesota; and the §Department of Radiology, Mayo Clinic, Rochester, Minnesota. The authors have reported that they have no relationships relevant to the contents of this paper to disclose. Presented as a poster and Rapid Fire Session at the annual meeting of the American Society of Echocardiography, on July 2, 2012, in Washington, DC.

Listen to this manuscript's audio summary by *JACC* Editor-in-Chief Dr. Valentin Fuster.

Manuscript received January 26, 2015; revised manuscript received March 11, 2015, accepted March 26, 2015.

echocardiography where the patient did not undergo excision.

SEE PAGE 2430

METHODS

This study was approved by the Mayo Clinic Institutional Review Board. The pathology and echocardiography databases of Mayo Clinic (Rochester, Minnesota) were queried for cases of PFE and cardiac myxoma occurring between January 1, 1995, and December 31, 2010, for retrospective review. Inclusion and exclusion criteria were applied (Figure 1). Patients were excluded from the study if any of the following was present:

- Patient declined access to their records for research.
- Inflammatory or infectious disease was present at time of incident PFE (n = 224).
- Transesophageal echocardiography (TEE) did not confirm findings of PFE suspected on transthoracic echocardiography (TTE) (n = 30) or, on review of images, PFE was not confirmed (n = 18).
- Surgeon could not confirm PFE or echocardiography-identified mass at surgery (n = 10) or histopathologic evaluation did not support the diagnosis (n = 33) (Table 1).

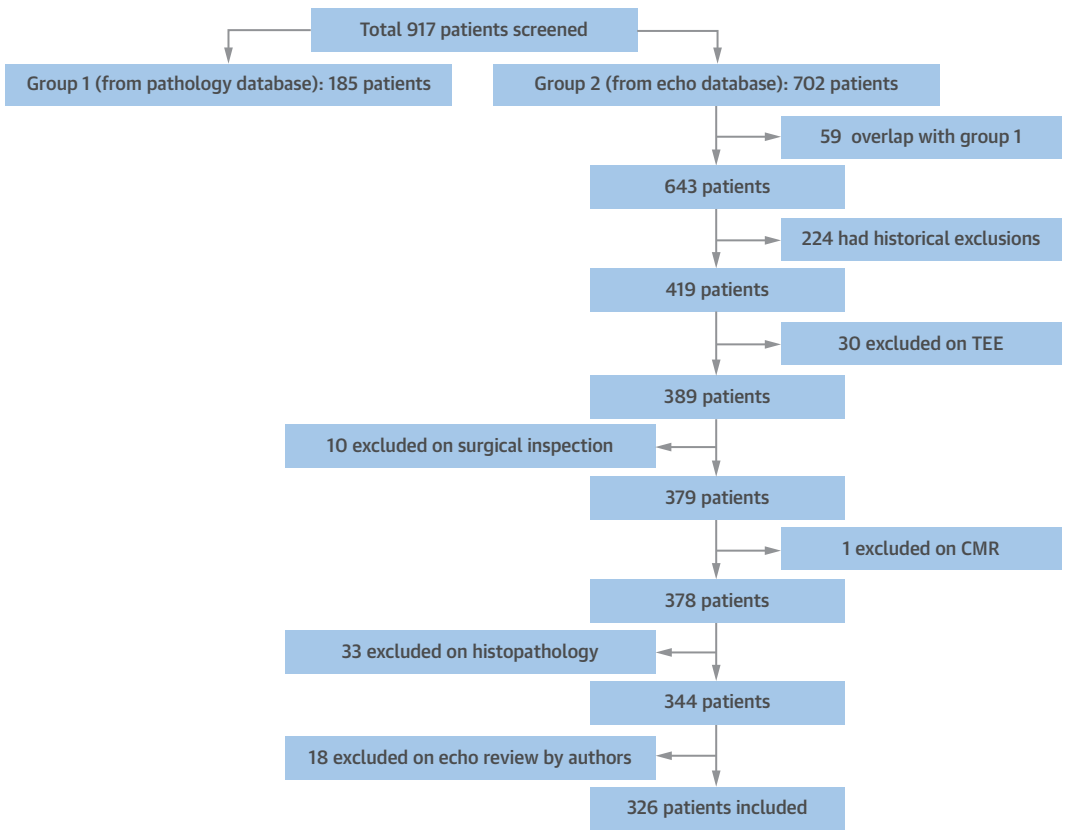
Patients were separated into 2 groups. Group 1 met histopathologic criteria for diagnosis of PFE. This group was divided into group 1a, including patients for whom PFE was the primary reason for surgical intervention, and group 1b, including patients for whom PFE excision was not the primary indication for surgery. Group 2 met echocardiographic criteria for PFE.

Histopathologic diagnosis was rendered by a cardiovascular pathologist (J.J.M. or W.D.E.) based

ABBREVIATIONS AND ACRONYMS

CI = confidence interval
CVA = cerebrovascular accident
HR = hazard ratio
IQR = interquartile range
NE = neurologic event(s)
PFE = papillary fibroelastoma(s)
RR = relative risk
TEE = transesophageal echocardiography
TIA = transient ischemic attack
TTE = transthoracic echocardiography

FIGURE 1 Study Flowchart



This chart outlines the inclusion and exclusion criteria for this study. CMR = cardiac magnetic resonance imaging; echo = echocardiography; TEE = transesophageal echocardiography.