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Pericardial/Myocardial Disease/Pulmonary Hypertension

PLATELET FUNCTION ANALYZER-100 (PFA-100) AS A POTENTIAL SCREENING TEST FOR OBSTRUCTIVE HYPERTROPHIC CARDIOMYOPATHY (HCM)

Moderated Poster Contributions
Poster Sessions, Expo North
Monday, March 11, 2013, 9:45 a.m.-10:30 a.m.

Session Title: What to Look for When Evaluating Hypertrophic Cardiomyopathy
Abstract Category: 23. Pericardial/Myocardial Disease
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Background: Screening for HCM is often recommended for first degree relatives of patients or in normal persons prior to competitive sports. PFA-100 is a proven screening test for congenital von Willebrand Disease. Abnormalities of von Willebrand factor (VWF) multimers were detected in 100% of patients with obstructive (O-HCM) (LeTourneau T, et al Circulation 2008;118:1550-1557) and 95 % of these had abnormal PFA-100.

Methods: We tested patients referred for echocardiography for HCM and controls (C). Groups compared included O-HCM, defined as resting peak gradient ≥30 mm Hg, latent HCM (L-HCM), peak gradient < 30 mm Hg but Valsalva-induced gradient ≥30 mm Hg, systolic anterior motion (SAM) with neither gradient ≥30 mm Hg, and patients post septal reduction therapy (SRT).

Results: PFA-100 Collagen-ADP (normal value < 121 seconds) results are displayed on 66 samples from 46 HCM patients; 39 had O-HCM, 13 had L-HCM, 4 had SAM, 11 post-SRT, and 44 C. In detecting O-HCM versus C, PFA-100 demonstrated sensitivity (95% CI) of 89% (74-97%) and specificity of 95% (83-99%), positive predictive value of 94% (80-99%), negative predictive value of 91% (78-97%), and a positive likelihood ratio of 19.7 (5.1-76.6).

Conclusion: We estimate that 95/100 relatives (prevalence 1:2) with a positive PFA-100 and 10/100 relatives with a negative test result will have O-HCM. For sports screening (prevalence 1:500) it is estimated that 4/100 patients with a positive test will have O-HCM while the risk with a negative test is minimal (2/10,000).