

**NEW CLASSIFICATION AND NATURAL HISTORY OF PATIENTS WITH
DIFFUSE PULMONARY ARTERIOVENOUS MALFORMATIONS: 29 YEAR
EXPERIENCE**

P Pierucci, J Murphy, K Henderson, JT Fahey, *D Chyun, RI White Jr.

Yale University School of Medicine, *Yale University School of Nursing, New Haven, CT, USA

Introduction: Patients with diffuse pulmonary arteriovenous malformations (PAVM), a small but important subset of the PAVM population, have significant morbidity and mortality.

Materials and Methods: 35 patients (20 female, 15 male) with diffuse PAVM, from a cohort of 821 consecutive patients with PAVM were evaluated during 2006. Diffuse PAVM were categorized angiographically: involvement of one or more segmental pulmonary arteries in one or both lungs. Hereditary hemorrhagic telangiectasia (HHT) status, gender, presence or absence of large (equal to or greater than 3mm diameter artery) focal PAVM, initial oxygen saturations and latest or current oxygen saturations post embolization, development of hemoptysis and survival were tabulated.

Results: HHT was present in 30/35 (86%) of the patients and diffuse PAVM were more commonly bilateral than unilateral. Focal PAVM were present in both groups but more commonly in those with bilateral involvement. All deaths occurred in those with bilateral involvement and were due to hemoptysis of bronchial artery origin (2), spontaneous liver necrosis (3), brain hemorrhage (1), brain abscess (1), and operative death during attempted lung transplant (1).

	UNILATERAL N=10	BILATERAL N=25	P-value
HHT	7 (70%)	23 (92%)	.11
GENDER			
MALE	8 (80%)	7 (28%)	.01
FEMALE	2 (20%)	18 (72%)	
FOCAL PAVMs	2 (20%)	16 (64%)	.02
INITIAL O ₂ SAT	.87±.07	.80±.08	.02
CURRENT O ₂ SAT	.91±.09	.84±.10	.08
HEMOPTYSIS	2 (20%)	6 (24%)	.34
DEATH	0	8 (32%)	.05

Conclusions: Patients with diffuse PAVM are a higher risk group for complications, than patients with focal PAVM and require yearly monitoring.