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Title: Bosentan for sarcoidosis associated pulmonary arterial hypertension (BoSAPH) was effective in advanced parenchymal lung disease

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Body: Introduction: We reported significant improvement in pulmonary artery mean (PAM) and pulmonary vascular resistance (PVR) for those treated with 16 weeks (wks) of bosentan (BOS) versus no change in those treated with placebo (PLA) for sarcoidosis associated pulmonary arterial hypertension (SAPH). Purpose of the study: To determine whether patients with advanced parenchymal disease were less likely to respond to BOS. Methods: Patients with SAPH confirmed by right heart catheterization (RHC) were randomized 2:1 to receive either BOS or PLA. After 16 wks of therapy, patients underwent repeat RHC. Patients had forced vital capacity (FVC) and Scadding chest x-ray (CXR) stage. Results: Of the 25 patients treated with BOS, 2 stopped study drug before wk 8. At 16 wks, 21had repeated RHC. Patients were subgrouped into those with FVC% ≥70% predicted (FVC≥70%) or below 70% (FVC≤70%) and those with CXR stage 0-3 (CXR<4) versus stage 4 (CXR=4). The table demonstrates the changes in PAM and PVR. There was significant improvement in PAM and PVR for those with FVC<70% but was not for those with FVC≥70%. No changes were seen for the PLA group.

PAM and PVR

	Number	Wk 0 PAM	Wk 16 PAM	Wk 0 PVR	Wk 16 PVR
FVC>70%	7	35+7.9	36+6.7	6.7+2.81	4.9+1.95
FVC<70%	14	36+6.6	32+10.0 *	5.5+3.01	4.1+2.08 *
CXR Stage 1-3	9	37+6.0	35+6.5	6.8+4.10	4.4+2.00 *
CXR Stage 4	12	35+7.6	30+10.1 *	5.1+1.34	4.4+2.14

Mean+S.D., * P<0.05

Conclusion: After 16 wks of treatment, bosentan therapy was associated with a significant improvement in PAM and PVR. These changes remained significant for patients with an FVC<70% predicted. SAPH patients with advanced parenchymal lung disease may respond to vasodilatory therapy.