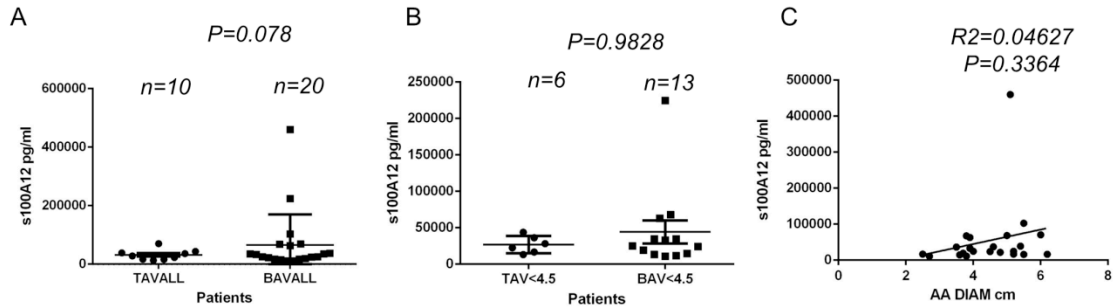


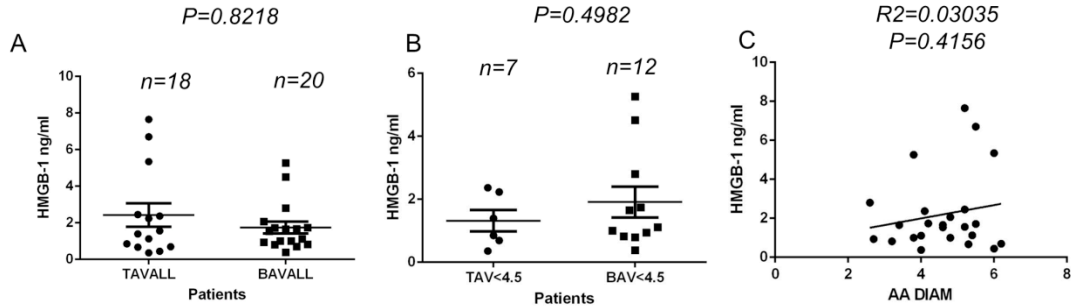
Supplemental Material

Supplemental Figure I



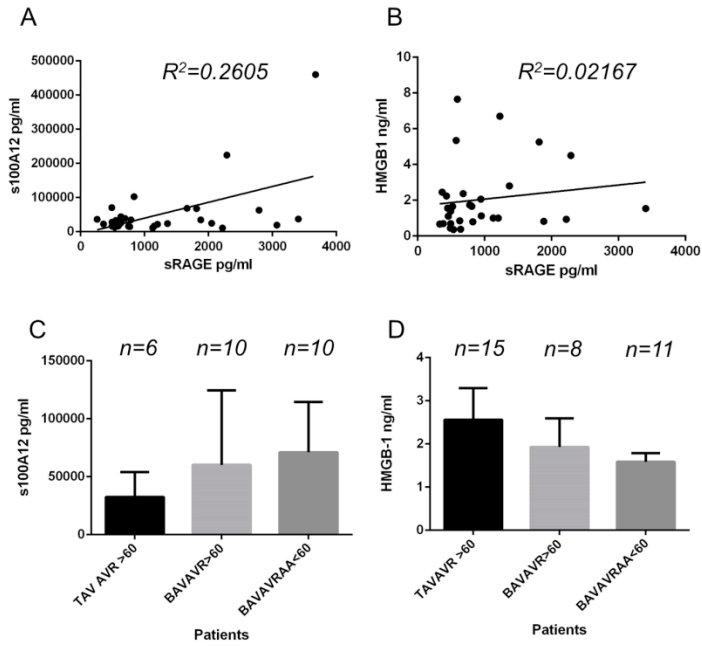
Circulating levels of s100A12 are not significantly different in BAV patients when compared to TAV. (A) sRAGE quantification of TAV (n=10) and BAV (n=20) patients. Dots represent values (pg/mL) from each patient \pm SEM. (B) sRAGE values in TAV (n=6) and BAV (n=13) patients meeting the criteria for surgical intervention (ascending aorta diameter <4.5 cm). (C) Linear regression to test the correlation between s100A12 plasma concentration and ascending aorta diameter.

Supplemental Figure II



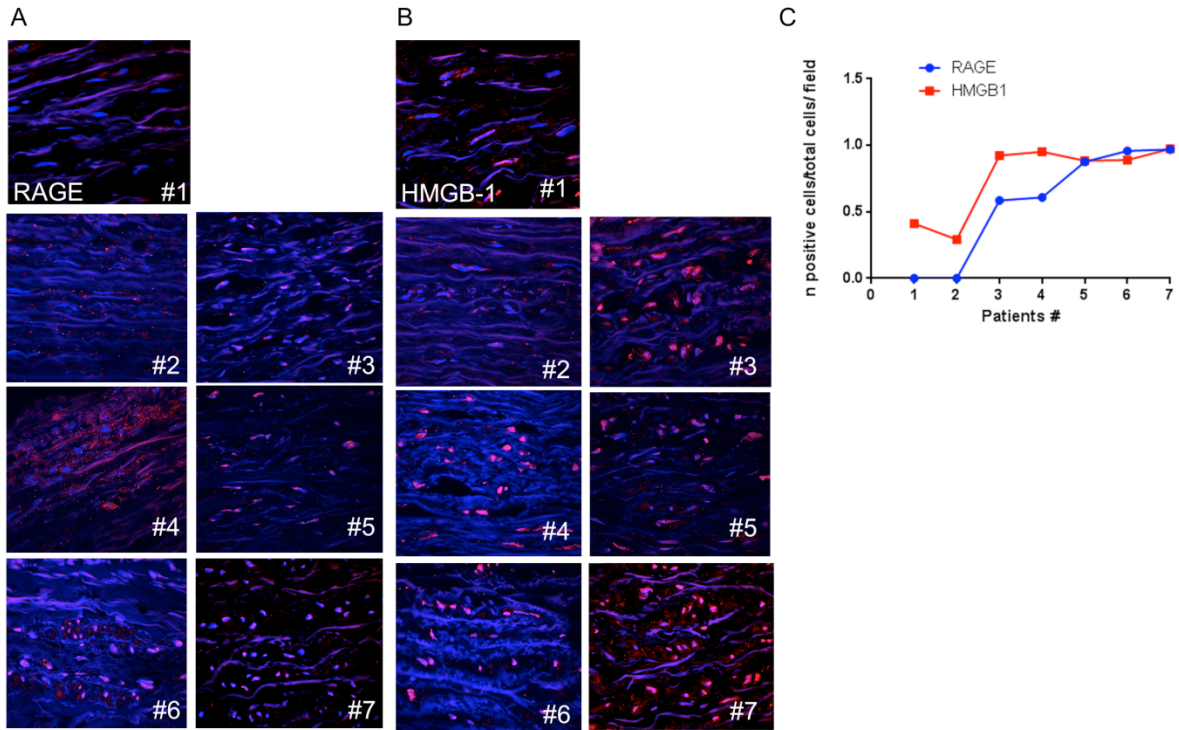
HMGB-1 plasma concentration is not significantly different in BAV patients when compared to TAV. (A) sRAGE quantification of TAV (n=18) and BAV (n=20) patients. Dots represent values (pg/mL) from each patient \pm SEM. (B) sRAGE values in TAV (n=7) and BAV (n=12) patients meeting the criteria for surgical intervention (ascending aorta diameter <4.5 cm). (C) Linear regression to test the correlation between HMGB-1 plasma concentration and ascending aorta diameter.

Supplemental Figure III



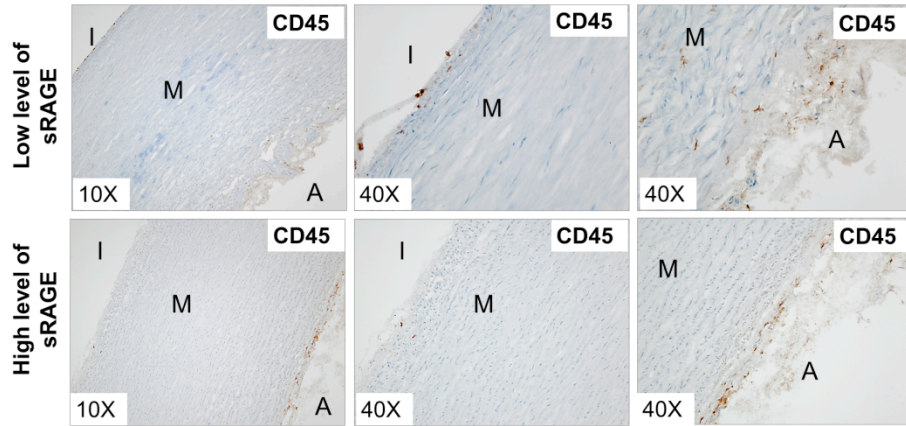
Absence of linear correlation between s100A12 and/or HMGB-1 and sRAGE level and distribution of sRAGE ligands in slow and fast progressors. (A-B) Linear regression to test the correlation between s100A12 or HMGB-1 plasma concentration and sRAGE circulating levels. (C) S100A12 and HMGB-1 distribution in patients defined as fast and slow progressor by sRAGE analysis.

Supplemental Figure IV



RAGE and HMGB-1 distribution in the ascending aorta of patients with low and high levels of circulating sRAGE. (A-B) Immunofluorescence analysis performed on ascending aorta sections obtained from patients #1-7. Red dots represents positive staining for either RAGE or HMGB-1. Confocal Images. Magnification 100X. (C) Graph representing RAGE and HMGB-1 positive staining quantification. Positive cells were counted in 4 field/section. Number of positive cells were averaged and plotted.

Supplemental Figure V



Few inflammatory infiltrates are detected in the aorta of patients with low and high levels of sRAGE.

Representative images of immunohistochemistry analysis for CD45 performed on ascending aorta sections obtained from patients with low and high levels of sRAGE. Magnification 10X and 40X.

I=Intima; M=Media;A=Adventitia.