Introduction
Kaposiform Hemangioendothelioma (KHE) is a rare, locally aggressive vascular tumor that typically occurs in early infancy. Lesions invade surrounding structures and rarely involute spontaneously. Mortality rate may be as high as 30%. KHE are distinct from other vascular tumors of infancy and are associated with both life threatening hemorrhage and Kasabach-Merritt Syndrome (KMS), a severe consumptive coagulopathy. Due to invasion of critical structures, surgery is often not possible. Medical treatment options include systemic steroids, antiplatelet drugs, and various chemotherapeutic agents. Propranolol has also been reported, but its full efficacy has yet to be established. Here, we report a case of a neonate with KHE complicated by pulmonary hypertension and KMS who was successfully managed with systemic steroids and propranolol.

Case Report
A full term male infant was born with a massive tumor of the head, neck, chest, and left upper extremity. The lesion was soft and was associated with several erythematous plaques and macules of varying sizes. Biopsy of one of these macules revealed a “benign angioma” with a “features of a kaposiform hemangioendothelioma, such as the discontinuous nature of the vascular lesion and the lack of GLUT1 staining.” Clinically, the patient was diagnosed with KHE.

Shortly after birth, the child developed KMS with thrombocytopenia and abnormal coagulation profiles. Platelets fell as low as 82x10^3/UL (normal 300-750x10^3/UL) with PT 17.9 seconds (normal 12-15.1), PTT 48.2 seconds (normal 24.7-37.1), Fibrinogen 428mg/dL (normal 211-423), and D-Dimer 2.57ug/mL FEU (normal <0.48).

The lesion also extended into the left chest and compressed both the left lung and mediastinal vasculature (see Figures 1 and 3). This compression resulted in pulmonary hypertension and acute cardiopulmonary failure. The patient was maintained on vascular pressors and begun on high dose corticosteroid therapy, prednisolone 3 mg/kg/day. This treatment stabilized the lesion and prevented further growth. Consideration was given to interferon and vincristine therapy, but the patient was felt to be too unstable. There was slow improvement in the lesion, and once the patient was off of vascular pressors, he was immediately begun on propranolol. Propranolol was quickly titrated to 2mg/kg/day, divided into thrice daily dosing with no alteration in hemodynamic status.

Within 2 weeks of beginning propranolol therapy, there was normalization of the patient’s coagulation profile, resolution of pulmonary hypertension, and marked clinical regression of the lesion (Figure 2).

Discussion
KHE are very rare tumors. As such, most of the literature dealing with these lesions consists of case reports. These lesions may be treated with a combination of vincristine, steroids, embolization, and surgery. Here, we report on a KHE complicated by cardiopulmonary compromise and KMS successfully managed with steroids and propranolol. In this case, propranolol seemed to be the key factor in resolution of the lesion. Steroid therapy provided control of the lesion, and induced a slow remission. However, initiation of propranolol resulted in rapid regression of the lesion and reversal of both the KMS and the pulmonary hypertension. To our knowledge, this case represents the fifth reported KHE involving the mediastinum and the second reported case of KHE with KMS successfully treated with propranolol. Hermans et al. reported on this treatment in 2011.

Conclusion
Propranolol may prove useful in the management of complicated KHE. The current body of reported cases could serve as the basis for further multi-institutional studies.

References