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LATE BREAKING Use of SANGUINATE™ in Acute Chest Syndrome

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Background

Acute Chest Syndrome (ACS) is defined as a new radiological density in chest x-ray associated with respiratory symptoms and hypoxemia. Regardless of the multiple possible triggering factors, the treatment is usually supportive with emphasis on improving oxygenation by simple or exchange transfusion. We describe a case of ACS in a patient with severe anemia who refused blood transfusion and was successfully treated with SANGUINATE.

Method (Patient)

23 year old female with sickle cell/ β^0 -thalassemia trait who was admitted to the MICU presenting with respiratory distress, PO₂ 54.3, sPO₂ 88.4%, pH 7.48 and PCO₂ 38.2 and anemia of 4.6. Over the last two years she has had approximately monthly sickle crises that did not result in severe complication. Patient had been recently discharged from another hospital after a 3-week stay for multi-organ failure, including NSTEMI (troponin 6.72) secondary to severe anemia, cerebral infarction, and stage 3 acute kidney injury with anuria requiring hemodialysis. Her lowest hemoglobin was 3.1 and was given IV iron and EPO with some improvement, but strictly refused transfusion.

Patient was intubated and begun on treatment for likely Acute Chest Syndrome, including IV vancomycin and cefepime. Hypertension (SBP >180) treated with IV labetalol. Oliguric acute tubular necrosis treated with hemodialysis; prescribed darbapoietin. Prior to intubation, patient was offered transfusions and despite risk of death, refused and requested to be supported in any other way possible.

500mL infusion of 40 mg/mL SANGUINATE™ was given intravenously and repeated in 24 hours. After initial extubation failure, two more units of SANGUINATE were provided to optimize for extubation. Response to treatment measured based upon her tolerance to weaning from the ventilator, clinical exam, bedside echo and Transcranial Doppler (TCD).

Results/Outcome

Patient was successfully extubated after second attempt, with no further need for re-intubation. Despite her persistently low Hb of 3.1 upon extubation, she reported improvement in her dyspnea. Her initial TCD revealed hyperemia with high blood flow velocities. Post-SANGUINATE the velocities gradually normalized. Her EF changed from 40% to 50%, PA changed from 57 to 63 and no significant change in RVSP or pericardial effusion over the 2 weeks course of ICU management.

Conclusion

SANGUINATE™ is a promising alternative to blood transfusion in treating ACS in SCD patients. It improved oxygenation and cerebral hemodynamics and facilitated extubation in the setting of severe anemia. Further research is needed to study its effect on transcranial flow and cardiac function.