

**Recent Advances in Acute
chest Syndrome in Sickle
Cell Disease**

Essay

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Introduction

Sickle cell disease is one of the most prevalent genetic diseases. Worldwide pulmonary disease , manifested as the acute chest syndrome (ACS) is a common complication of Sickle cell disease. Accounting for 25% of premature deaths.

The last decade has witnessed a convergence of research pathways that were leading towards a better understanding of the new possible pathophysiology and therapies for the disease and new data on the effect of nitric oxide (NO) on sickle cell hemoglobin and interaction between them. (Bunn, 1994)

The acute chest syndrome is a common form of lung injury in sickle cell disease. When severe, this syndrome is analogous to the acute respiratory distress syndrome . The acute chest syndrome is the second common cause of hospitalization among patients with sickle cell disease and the leading cause of admission to an intensive care unit and premature death.

It is found that about 60% of these patients with severe acute chest syndrome (ACS) had pulmonary hypertension and cor pulmonale. The risk for developing an ACS episode appears to be increased following surgery, with an average to the development of ACS of 3 days post-surgery. (Gladwin and Vichinsky, 2008)

The perioperative period can offer a unique insight into the origins of acute and chronic complications of sickle cell disease. An examination of the assumptions and consequences of anesthetic practice aimed at the prevention and treatment of these complications, similarly can provide a useful distillation of management principles.

(Firth and Head, 2004)