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# Effect of hydroxyurea in preventing ocular complications in Sickle Cell Disease (Italian Study)

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## Abstract

**Purpose:** Sickle cell disease (SCD) is a life-threatening genetic disorder which leads to most sever retinal manifestations requiring immediate medical attention. The ocular changes that compromise the vision of patients with SCD are the consequences of a complex systemic pathophysiological process. The prevention of these ocular complications can be achieved through the use of new drugs that focus on the physiopathology of the disease. Two disease-modifying therapies, hydroxyurea and long-term blood transfusions, are available but underused. We have investigated the prevention effect of hydroxyurea on ocular complications in Italian patients with SCD. <br />

**Methods:** We studied 123 patients with SCD including 70 treated with hydroxyurea and 53 with other therapy.<br />

**Results:** Among all patients studied, 16 patients (13%) developed retinopathy. In the group treated with hydroxyurea(70 patients), 4 patients (5,71%) developed ocular complications. In the second group (53 patients) without hydroxyurea treatment, 12 patients (22,64%) developed ocular manifestations of SCD. Independent of hydroxyurea, patients with a HbF <15% had 7.1-fold (95% confidence interval, 1.5-33.6) higher odds of developing retinopathy. In patients treated with hydroxyurea, those with retinopathy had lower HbF levels compared to patients without retinopathy (9% vs. 16%; P = 0.005). <br />

**Conclusions:** We report a protective benefit of hydroxyurea treatment regarding ocular complications throught high levels of HbF. <br />