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Analysis of Sickle Cell Disease and Maternal Mortality in United Kingdom

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Abstract: Aims and Objectives: To determine the incidence of maternal mortality amongst pregnant women with sickle cell disease (SCD) in the United Kingdom and to determine exact cause of death in these women. Background: SCD is caused by the 'sickle' gene and is characterized by episodes of severe bone pain and other complications like acute chest syndrome, chronic pulmonary hypertension, stroke, retinopathy, chronic renal failure, hepato-splenic crises, avascular bone necrosis, sepsis and leg ulcers. SCD is a continual cause of maternal mortality and fetal complications, and it comprises 1.5% of all Direct and Indirect deaths in the UK. Sepsis following premature rupture of membranes with ascending infection, post-partum infection and pre-labour overwhelming septic shock is one of its leading causes of death. Over the last fifty years of maternal mortality reports in UK, between 1 to 4 pregnant women died in each triennium. Material and Method: This is a retrospective study that involves pregnant women who died from SCD complications in the UK between 1952-2012. Data were collected from the UK Confidential Enquiries into Maternal Death and its causes between 1952-2012. Prior to 1985, exact cause of death in this cohort was not recorded. Results: 33 deaths reported between 1964 and 1984. 17 deaths were reported due to sickle cell disease between 1985 and 2012. Five women in this group died of sickle cell crisis, one woman had liver sequestration crisis, two women died of venous thromboembolism, two had myocardial fibrosis and three died of sepsis. Remaining women died of amniotic fluid embolism, SUDEP, myocardial ischemia and intracranial haemorrhage. Conclusion: The leading causes of death in sickle cell sick pregnant women are sickle cell crises, sepsis, venous thrombosis and thromboembolism. Prenatal care for women with SCD should be managed by a multidisciplinary team that includes an obstetrician, nutritionist, primary care physician, and haematologist. In every sick Sickle Cell woman Sickle Cell crises should be on the top of the list of differential diagnosis. Aggressive treatment of complications with low threshold to commence broad-spectrum antibiotics and LMWH contribute to better outcomes.

Keywords: incidence, maternal mortality, sickle cell disease (SCD), uk

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