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[Intervention Review]

Interventions for treating intrahepatic cholestasis in people with sickle cell disease

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ABSTRACT

Background

Sickle cell disease is the most common hemoglobinopathy occurring worldwide and sickle cell intrahepatic cholestasis is a complication long recognized in this population. Cholestatic liver diseases are characterized by impaired formation or excretion (or both) of bile from the liver. There is a need to assess the clinical benefits and harms of the interventions used to treat intrahepatic cholestasis in people with sickle cell disease. This is an update of a previously published Cochrane Review.

Objectives

To assess the benefits and harms of the interventions for treating intrahepatic cholestasis in people with sickle cell disease.

Search methods

We searched the Cystic Fibrosis and Genetic Disorders Group's Haemoglobinopathies Trials Register, which comprises references identified from comprehensive electronic database searches and handsearching of relevant journals and abstract books of conference proceedings. We also searched the LILACS database (1982 to 23 May 2017), the WHO International Clinical Trials Registry Platform Search Portal (23 May 2017) and ClinicalTrials.gov.

Date of last search of the Cochrane Cystic Fibrosis and Genetic Disorders Group's Haemoglobinopathies Trials Register: 12 April 2017.

Selection criteria

We searched for published or unpublished randomised controlled trials.

Data collection and analysis

Each author intended to independently extract data and assess the risk of bias of the trials by standard Cochrane methodologies; however, no trials were included in the review.

Main results

There were no randomised controlled trials identified.

Authors' conclusions

This updated Cochrane Review did not identify any randomised controlled trials assessing interventions for treating intrahepatic cholestasis in people with sickle cell disease. Randomised controlled trials are needed to establish the optimum treatment for this condition.

PLAIN LANGUAGE SUMMARY

Interventions for treating intrahepatic cholestasis in people with sickle cell disease

Review question

We aimed to review the evidence for treating intrahepatic cholestasis (liver diseases where bile is either not formed or excreted properly (or both)) in people with sickle cell disease. This is an update of a previously published Cochrane Review.

Background

Sickle cell disease is an inherited condition and the most common hemoglobinopathy occurring worldwide. In 2006, the WHO declared sickle cell disease as a major public health problem with an estimated 70% of sufferers living in Africa. It is common among people with sub-Saharan African, Indian, Middle Eastern or Mediterranean ancestry.

Sickle cell disease affects the hepatobiliary system (liver, gall bladder, bile ducts), with repeated reduced blood flow and the formation of bilirubin stones (a type of gallstone). This results from sickled cells blocking blood vessels and from a reduction in the lifespan of these cells. Sickle cell intrahepatic cholestasis or sickle cell hepatopathy (abnormal or diseased liver) is one complication of the disease. When diagnosed, people with this condition may show an intense yellowing of skin and eyes due to an increasing level of serum bilirubin (jaundice), tiredness, generalized itching, increasing pain in the upper right quadrant of the abdomen due to gallstones, an enlarged liver and lower hemoglobin levels.

Sickle cell intrahepatic cholestasis, while uncommon, is a potentially fatal complication with a high death rate. There is no agreement in how to diagnose or treat this condition. We wanted to assess the benefits and harms of treatments for intrahepatic cholestasis in people with sickle cell disease.

Search date

The evidence is current to 12 April 2017.

Study characteristics

We were not able to find any eligible trials.

Key results

No trials were identified.

Quality of the evidence

There is no randomised controlled trial evidence of any intervention for treating intrahepatic cholestasis in people with sickle cell disease. Trials are needed to establish the best treatment for this condition.