

Cochrane Database of Systematic Reviews - - Cochrane Review

Ivacaftor-tezacaftor-elexacaftor, tezacaftor-ivacaftor and lumacaftor-ivacaftor for treating cystic fibrosis: a systematic review and economic evaluation.

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Study design (if review, criteria of inclusion for studies)

Randomised controlled trials (RCTs) and quasi-RCTs.

Participants

Children and adults of all ages and either gender, diagnosed with CFLD with cirrhosis or liver failure, PHT or variceal bleeding (or both).

Interventions

1. Pharmacological interventions (e.g. non-selective betablockers) for any duration compared to placebo or no intervention 2. Endoscopic interventions (e.g. band ligation, sclerotherapy) compared to active control 3. TIPSS compared to active control 4. Surgical interventions (e.g. surgical PS shunt, liver transplantation) compared to active control

Outcome measures

Primary outcomes 1. Change in variceal bleeding and portal pressure i) development of first bleeding episode (primary prevention) ii) re-bleeding following endoscopic treatment (secondary prevention) 2. Adverse effects i) of non-surgical interventions (e.g. aggravated congestive cardiac failure, respiratory distress, hypotension from non-selective beta-blockers, stenosis, occlusion, thrombosis, worsened encephalopathy after TIPSS, perforation of oesophagus, oesophageal ulceration, oesophageal stricture after sclerotherapy) ii) of surgical interventions (e.g. thrombosis, occlusion, onset or worsening of encephalopathy, infection, rejection) Secondary outcomes 1. Nutritional status 2. Respiratory outcomes.

Main results

A comprehensive search of the literature did not identify any published eligible randomised controlled trials.

Authors' conclusions

In order to develop the best source of evidence, there is a need to undertake randomised controlled trials of interventions for preventing and managing advanced liver disease in adults and children with cystic fibrosis.

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See also

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Keywords

Liver diseases; non pharmacological intervention - surg; pharmacological_intervention;