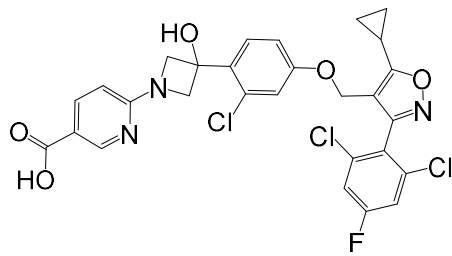


CLAIMS

1. A compound having the following Formula (I):



or a pharmaceutically acceptable salt thereof.

2. The compound according to claim 1 or pharmaceutically acceptable salt thereof for use in the treatment of a Farnesoid X Receptor (FXR) mediated condition,

wherein the FXR mediated condition is selected from the group consisting of:

a chronic intrahepatic or extrahepatic cholestatic condition;

liver fibrosis;

a chronic or obstructive inflammatory disorder of the liver;

liver cirrhosis;

liver steatosis or an associated syndrome;

a cholestatic or fibrotic effect that is associated with alcohol-induced cirrhosis or with a viral-borne form of hepatitis;

acute or chronic liver failure;

liver ischemia after major liver resection;

chemotherapy associated steatohepatitis (CASH);

Primary Biliary Cirrhosis (PBC);

Primary Sclerosing Cholangitis (PSC);

a neoplastic disease of the gastrointestinal tract or liver; and

an Inflammatory Bowel Disease (IBD);

a lipid disorder or lipoprotein disorder;

Type I Diabetes;

Type II Diabetes;

clinical complications of Type I and Type II Diabetes selected from the group consisting of diabetic nephropathy, diabetic neuropathy, diabetic retinopathy and other observed effects of clinically manifest long term Diabetes;

Non-Alcoholic Fatty Liver Disease (NAFLD);

Non-Alcoholic Steatohepatitis (NASH);

obesity;

a metabolic syndrome selected from the group consisting of combined conditions of dyslipidemia, diabetes and abnormally high body-mass index;

acute myocardial infarction;

acute stroke; and

thrombosis that occurs as an endpoint of chronic obstructive atherosclerosis;

a non-malignant hyperproliferative disorder;

a malignant hyperproliferative disorder selected from the group consisting of hepatocellular carcinoma, colon adenoma, and polyposis;

colon adenocarcinoma;

breast cancer;

pancreas adenocarcinoma; and

Barrett's esophagus.

3. The compound for use according to claim 2 or pharmaceutically acceptable salt thereof wherein the FXR mediated condition is a liver disease.

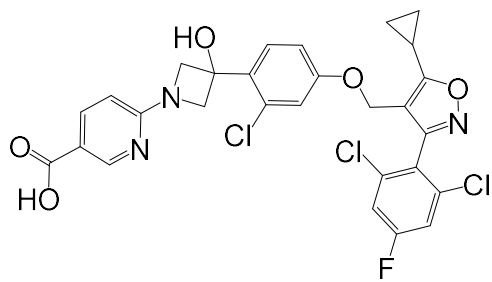
4. The compound for use according to claim 2 or 3 or pharmaceutically acceptable salt thereof, wherein the FXR mediated condition is Non-Alcoholic Steatohepatitis (NASH).

5. The compound for use according to claim 2 or 3 or pharmaceutically acceptable salt thereof, wherein the FXR mediated condition is Primary Sclerosing Cholangitis (PSC).

6. The compound for use according to claims 2 or 3 or pharmaceutically acceptable salt thereof, wherein the FXR mediated condition is Primary Biliary Cholangitis (PBC).

7. The compound for use according to claim 2 or 3 or pharmaceutically acceptable salt thereof, wherein the FXR mediated condition is liver fibrosis.

8. A pharmaceutical composition comprising a compound having the following Formula (I):



or a pharmaceutically acceptable salt thereof.

9. The pharmaceutical composition of claim 8, further comprising a pharmaceutically acceptable carrier.

10. The use of a compound or pharmaceutically acceptable salt of claim 1, or a pharmaceutical composition of claim 8 or 9 in the manufacture of a medicament for treating a patient having a Farnesoid X Receptor (FXR) mediated condition,

wherein the FXR mediated condition is selected from the group consisting of:

a chronic intrahepatic or extrahepatic cholestatic condition;

liver fibrosis;

a chronic or obstructive inflammatory disorder of the liver;

liver cirrhosis;

liver steatosis or an associated syndrome;

a cholestatic or fibrotic effect that is associated with alcohol-induced cirrhosis or with a viral-borne form of hepatitis;

acute or chronic liver failure;

liver ischemia after major liver resection;

chemotherapy associated steatohepatitis (CASH);

Primary Biliary Cirrhosis (PBC);

Primary Sclerosing Cholangitis (PSC);

a neoplastic disease of the gastrointestinal tract or liver; and

an Inflammatory Bowel Disease (IBD);

a lipid disorder or lipoprotein disorder;

Type I Diabetes;

Type II Diabetes;

clinical complications of Type I and Type II Diabetes selected from the group consisting of diabetic nephropathy, diabetic neuropathy, diabetic retinopathy and other observed effects of clinically manifest long term Diabetes;

Non-Alcoholic Fatty Liver Disease (NAFLD);

Non-Alcoholic Steatohepatitis (NASH);

obesity;

a metabolic syndrome selected from the group consisting of combined conditions of dyslipidemia, diabetes and abnormally high body-mass index;

acute myocardial infarction;

acute stroke; and

thrombosis that occurs as an endpoint of chronic obstructive atherosclerosis;

a non-malignant hyperproliferative disorder;

a malignant hyperproliferative disorder selected from the group consisting of hepatocellular carcinoma, colon adenoma, and polyposis;

colon adenocarcinoma;

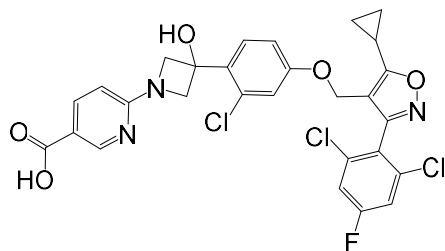
breast cancer;

pancreas adenocarcinoma; and

Barrett's esophagus.

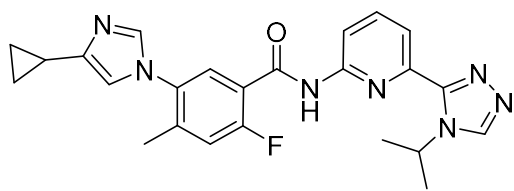
11. The use of a therapeutically effective amount of an Apoptosis Signal-Regulating Kinase 1 (ASK1) inhibitor in combination with a therapeutically effective amount of a Farnesoid X Receptor (FXR) agonist, in the manufacture of a medicament for the treatment/prevention of a liver disease in a patient in need thereof

wherein the FXR agonist is a compound of Formula (I):



or a pharmaceutically acceptable salt thereof.

12. The use of claim 11, wherein the ASK1 inhibitor is a compound of Formula (II):



or a pharmaceutically acceptable salt, a stereoisomer, a mixture of stereoisomers, or a tautomer thereof.

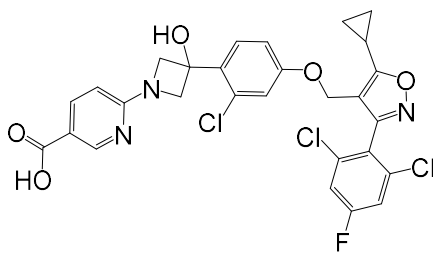
13. The use of claim 11 to 12, wherein FXR agonist and the ASK1 inhibitor are to be administered separately.

14. The use of any one of claims 11 to 13, wherein the liver disease is non-alcoholic steatohepatitis (NASH).

15. The use of any one of claims 11 to 13, wherein the liver disease is Primary Sclerosing Cholangitis (PSC).

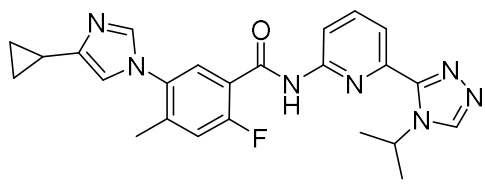
16. The use of any one of claims 11 to 13, wherein the liver disease is Primary Biliary Cirrhosis (PBC).

17. A pharmaceutical composition comprising a therapeutically effective amount of an Apoptosis Signal-Regulating Kinase 1 (ASK1) inhibitor and a therapeutically effective amount of a Farnesoid X Receptor (FXR) agonist, wherein the FXR agonist is a compound of Formula (I):



or a pharmaceutically acceptable salt thereof.

18. The pharmaceutical composition of claim 17, wherein the ASK1 inhibitor is a compound of Formula (II):

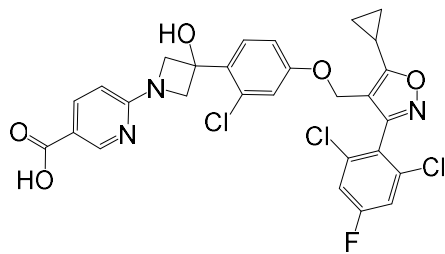


or a pharmaceutically acceptable salt, a stereoisomer, a mixture of stereoisomers, or a tautomer thereof.

19. The pharmaceutical composition of claim 17 or 18, further comprising a pharmaceutically acceptable carrier.

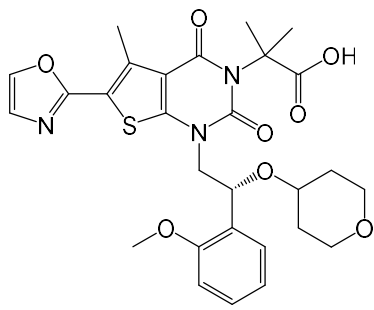
20. The use of a therapeutically effective amount of an Acetyl CoA Carboxylase (ACC) inhibitor in combination with a therapeutically effective amount of a Farnesoid X Receptor (FXR) agonist, in the manufacture of a medicament for treating and/or preventing a liver disease in a patient in need thereof

wherein the FXR agonist is a compound of Formula (I):



or a pharmaceutically acceptable salt thereof.

21. The use of claim 20, wherein the ACC inhibitor is a compound of Formula (III):



or a pharmaceutically acceptable salt, a stereoisomer, a mixture of stereoisomers, or a tautomer thereof.

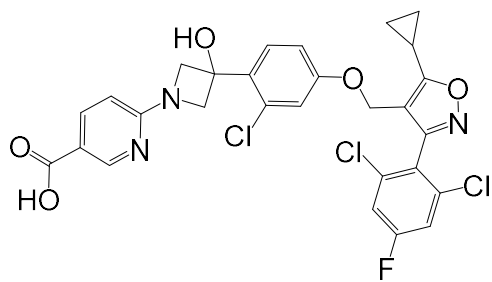
22. The use of claim 20 or 21, wherein the ACC inhibitor and the FXR agonist are administered separately.

23. The use of any one of claims 20 to 22, wherein the liver disease is non-alcoholic steatohepatitis (NASH).

24. The use of any one of claims 20 to 22, wherein the liver disease is Primary Sclerosing Cholangitis (PSC).

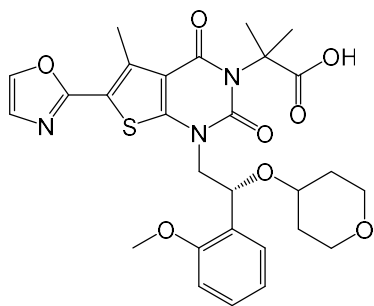
25. The use of any one of claims 20 to 22, wherein the liver disease is Primary Biliary Cirrhosis (PBC).

26. A pharmaceutical composition comprising a therapeutically effective amount of an Acetyl CoA Carboxylase (ACC) inhibitor and a therapeutically effective amount of a Farnesoid X Receptor (FXR) agonist, wherein the FXR agonist is a compound of Formula (I):



or a pharmaceutically acceptable salt thereof.

27. The pharmaceutical composition of claim 26, wherein the ACC inhibitor is a compound of Formula (III):

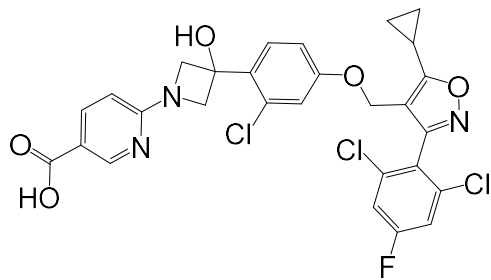


or a pharmaceutically acceptable salt, a stereoisomer, a mixture of stereoisomers, or a tautomer thereof.

28. The pharmaceutical composition of claim 26 or 27, further comprising a pharmaceutically acceptable carrier.

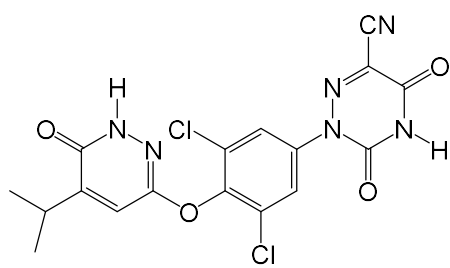
29. The use of a therapeutically effective amount of a Thyroid Hormone Receptor (THR) β agonist in combination with a therapeutically effective amount of a Farnesoid X Receptor (FXR) agonist, in the manufacture of a medicament for treating and/or preventing a liver disease in a patient in need thereof.

wherein the FXR agonist is a compound of Formula (I):



or a pharmaceutically acceptable salt thereof.

30. The use of claim 29, wherein the THR β agonist is a compound of Formula (IV):



or a pharmaceutically acceptable salt, a stereoisomer, a mixture of stereoisomers, or a tautomer thereof.

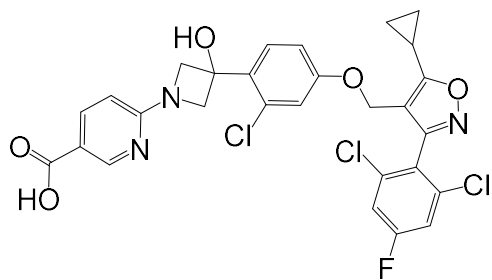
31. The use of claim 29 or 30, wherein the THR β agonist and the FXR agonist are administered separately.

32. The use of any one of claims 29 to 31, wherein the liver disease is non-alcoholic steatohepatitis (NASH).

33. The use of any one of claims 29 to 31, wherein the liver disease is Primary Sclerosing Cholangitis (PSC).

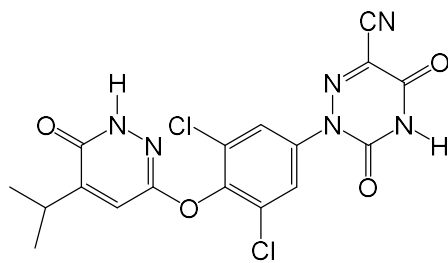
34. The use of any one of claims 29 to 31, wherein the liver disease is Primary Biliary Cirrhosis (PBC).

35. A pharmaceutical composition comprising a therapeutically effective amount of a Thyroid Hormone Receptor (THR) β and a therapeutically effective amount of a Farnesoid X Receptor (FXR) agonist, wherein the FXR agonist is a compound of Formula (I):



or a pharmaceutically acceptable salt thereof.

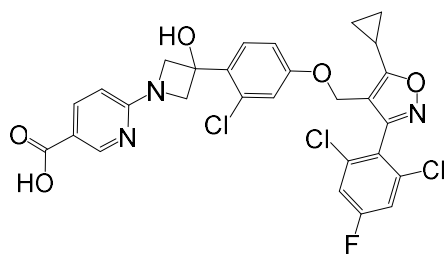
36. The pharmaceutical composition of claim 35, wherein the THR β agonist is a compound of Formula (IV):



or a pharmaceutically acceptable salt, a stereoisomer, a mixture of stereoisomers, or a tautomer thereof.

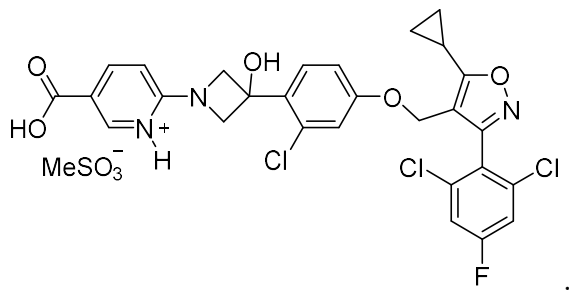
37. The pharmaceutical composition of claim 35 or 36, further comprising a pharmaceutically acceptable carrier.

38. Use of a therapeutically effective amount of a compound of Formula (I):



or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for inhibiting UGT1A1 in a patient having a liver disease.

39. A crystalline form of a compound of the following formula:



40. The crystalline form of claim 39, wherein the crystalline form is Form I.
41. The crystalline form of claim 40, characterized by an X-ray diffraction pattern having 2 θ -reflections at 9.6, 19.3, and 22.6 degrees 2 θ , plus or minus 0.2 degrees 2 θ .
42. The crystalline form of claim 40 or 41, characterized by an X-ray diffraction pattern further comprising 2 θ -reflections at 3.2, 6.4, and 12.8 degrees 2 θ , plus or minus 0.2 degrees 2 θ .
43. The crystalline form of any of claims 40-42, characterized by an X-ray diffraction pattern further comprising 2 θ -reflections at 22.1, 25.8, 29.1 degrees 2 θ , plus or minus 0.2 degrees 2 θ .
44. The crystalline form of any of claims 40-43, having a differential scanning calorimetry thermogram comprising an endothermic with onset at about 221 °C.