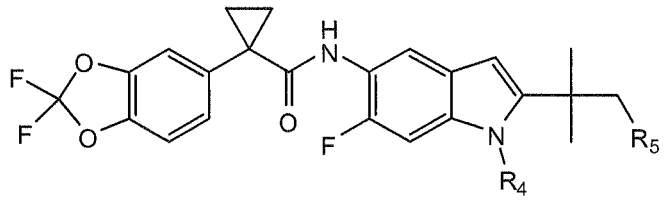
We claim:

1. A compound of formula IIa:



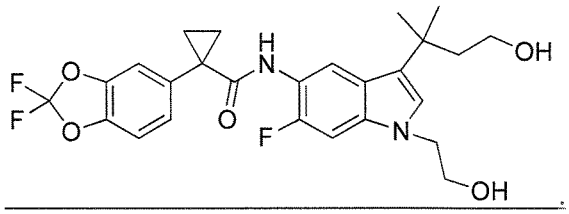
IIa

or a pharmaceutically acceptable salt thereof, wherein independently for each occurrence:

R_4 is H, $-CH_2CH(OH)CH_2OH$, $-CH_2CH_2N^+(CH_3)_3$, or $-CH_2CH_2OH$; and

R_5 is OH, $-CH_2OCH_2CH(OH)CH_2OH$, $-CH_2OH$, or R_4 and R_5 taken together form a fused pyrrolidine ring;

provided that the compound is not

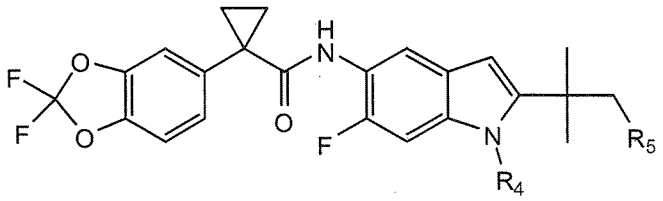


2. The compound of claim 1, wherein R_4 is (*R*)-$CH_2CH(OH)CH_2OH$, (*S*)-$CH_2CH(OH)CH_2OH$, $-CH_2CH_2N^+(CH_3)_3$, or $-CH_2CH_2OH$.

3. The compound of claim 1, wherein R_5 is OH, $-CH_2OCH_2CH(OH)CH_2OH$, or $-CH_2OH$.

4. The compound of claim 1, wherein R_4 is (*R*)-$CH_2CH(OH)CH_2OH$, (*S*)-$CH_2CH(OH)CH_2OH$, $-CH_2CH_2N^+(CH_3)_3$, or $-CH_2CH_2OH$; and R_5 is OH, $-CH_2OCH_2CH(OH)CH_2OH$, or $-CH_2OH$.

5. The compound of claim 1 which is selected from:



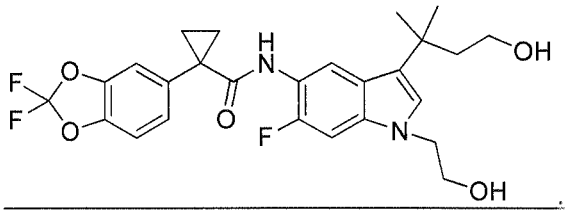
IIa

or a pharmaceutically acceptable salt thereof, wherein independently for each occurrence:

R_4 is H, $-CH_2CH(OH)CH_2OH$, $-CH_2CH_2N^+(CH_3)_3$, or $-CH_2CH_2OH$; and

R_5 is OH, $-CH_2OCH_2CH(OH)CH_2OH$, $-CH_2OH$, or R_4 and R_5 taken together form a fused pyrrolidine ring;

provided that the compound is not

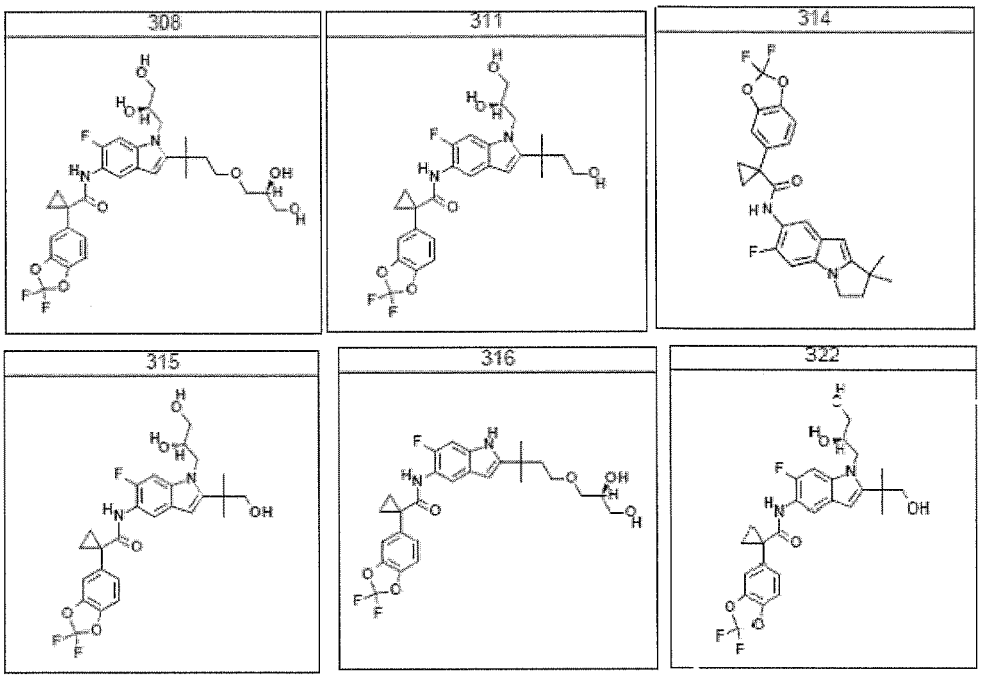


9. The in vitro use of claim 8, wherein R_4 is (*R*)-$CH_2CH(OH)CH_2OH$, (*S*)-$CH_2CH(OH)CH_2OH$, $-CH_2CH_2N^+(CH_3)_3$, or $-CH_2CH_2OH$.

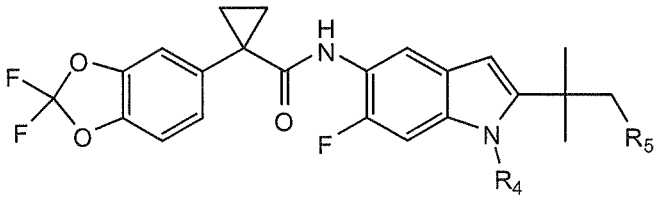
10. The in vitro use of claim 8, wherein R_5 is OH, $-CH_2OCH_2CH(OH)CH_2OH$, or $-CH_2OH$.

11. The in vitro use of claim 8, wherein R_4 is (*R*)-$CH_2CH(OH)CH_2OH$, (*S*)-$CH_2CH(OH)CH_2OH$, $-CH_2CH_2N^+(CH_3)_3$, or $-CH_2CH_2OH$; and R_5 is OH, $-CH_2OCH_2CH(OH)CH_2OH$, or $-CH_2OH$.

12. The in vitro use of claim 8, wherein the compound is selected from:



13. The compound having formula IIa ~~wherein~~:



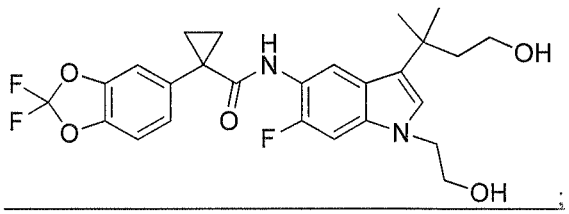
IIa

or a pharmaceutically acceptable salt thereof, wherein independently for each occurrence:

R_4 is H, $-CH_2CH(OH)CH_2OH$, $-CH_2CH_2N^+(CH_3)_3$, or $-CH_2CH_2OH$; and

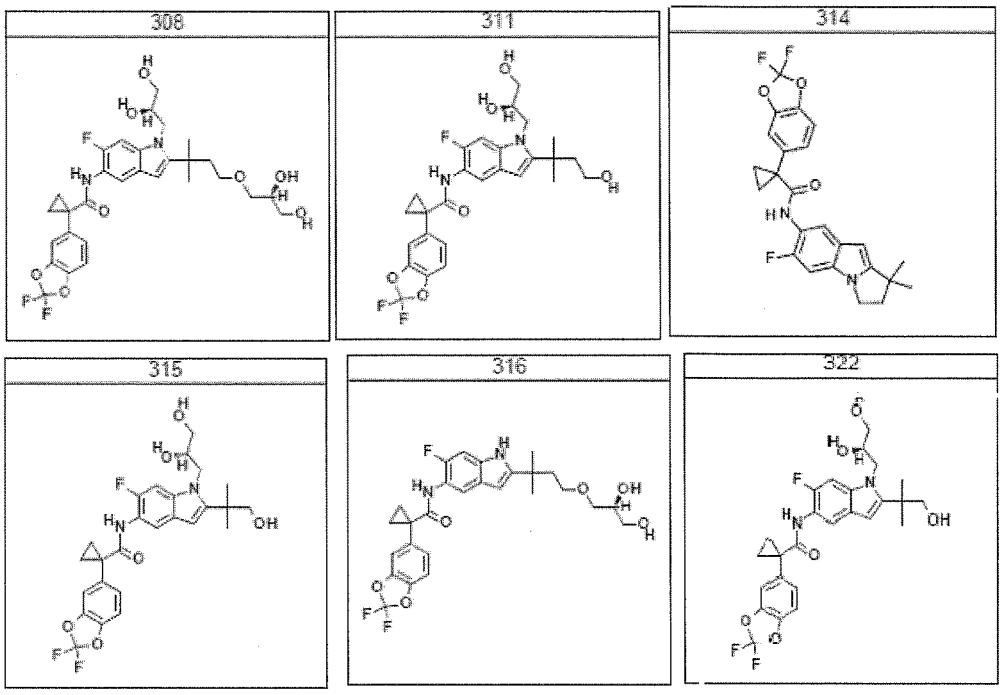
R_5 is OH, $-CH_2OCH_2CH(OH)CH_2OH$, $-CH_2OH$, or R_4 and R_5 taken together form a fused pyrrolidine ring;

provided that the compound is not



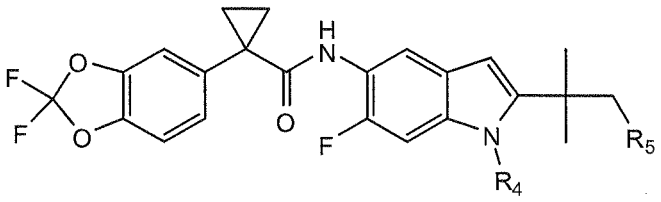
;

for use in treating a condition, disease, or disorder selected from cystic fibrosis, hereditary emphysema, hereditary hemochromatosis, coagulation-fibrinolysis deficiencies, such as protein C deficiency, Type 1 hereditary angioedema, lipid



18. A kit for use in measuring the activity of a ABC transporter o · a fragment thereof in a biological sample *in vitro* or *in vivo*, comprising:

(i) a first composition comprising a compound of formula II**a**:



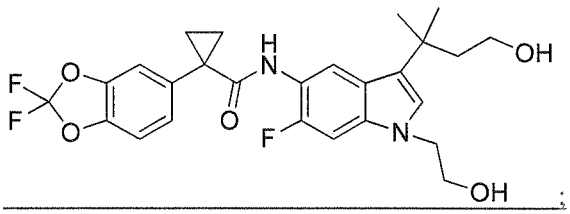
IIa

or a pharmaceutically acceptable salt thereof, wherein independently for each occurrence:

R_4 is H, $-CH_2CH(OH)CH_2OH$, $-CH_2CH_2N^+(CH_3)_3$, or $-CH_2CH_2OH$; and

R_5 is OH, $-CH_2OCH_2CH(OH)CH_2OH$, $-CH_2OH$, or R_4 and R_5 taken together form a fused pyrrolidine ring;

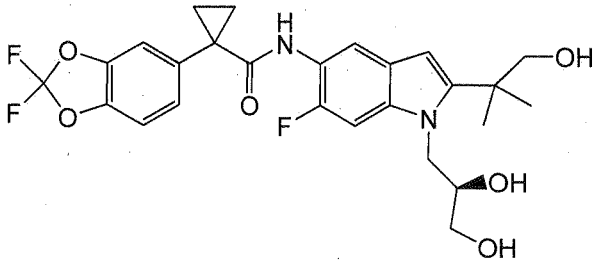
provided that the compound is not



;

(ii) instructions for:

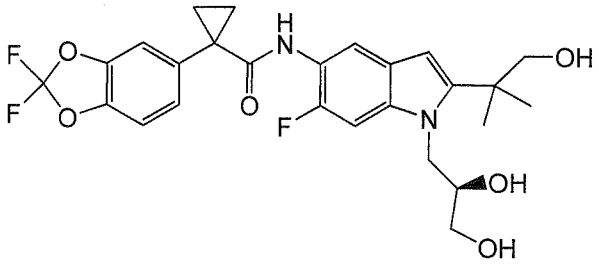
23. A compound ~~of the formula~~which is:



or a pharmaceutically acceptable salt thereof.

24. A pharmaceutical composition comprising

(i) a compound which is:~~of the formula;~~



(ii) a pharmaceutically acceptable carrier.

25. The composition of claim 24, further comprising an additional agent wherein the additional agent is N-(5-hydroxy-2,4-ditert-butyl-phenyl)-4-oxo-1H-quinoline-3-carboxamide.

26. Use of a compound of claim 23 in the manufacture of a medicament for the treatment of a disease or disorder selected from male infertility and pancreatic insufficiency.

27. Use of a compound of claim 23 in the manufacture of a medicament for the treatment of cystic fibrosis.

28. The composition of claim 6, further comprising an additional agent wherein the additional agent is N-(5-hydroxy-2,4-ditert-butyl-phenyl)-4-oxo-1H-quinoline-3-carboxamide.

29. Use of a compound of claim 23 in the manufacture of a medicament for the treatment of a disease or disorder selected from hereditary emphysema, COPD, and dry-eye disease.