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(54) Title **MODULATORS OF CYSTIC FIBROSIS TRANSMEMBRANE CONDUCTANCE REGULATOR**

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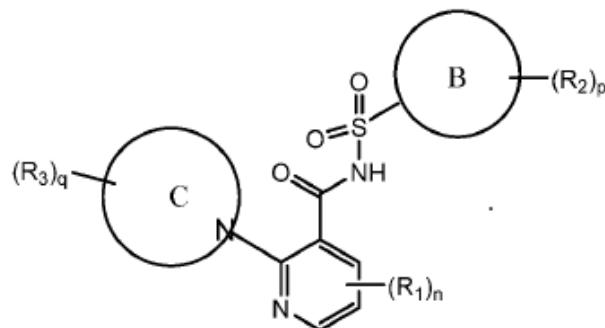
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Enclosed is a translation of the patent claims in Norwegian. Please note that as per the Norwegian Patents Acts, section 66i the patent will receive protection in Norway only as far as there is agreement between the translation and the language of the application/patent granted at the EPO. In matters concerning the validity of the patent, language of the application/patent granted at the EPO will be used as the basis for the decision. The patent documents published by the EPO are available through Espacenet (<http://worldwide.espacenet.com>) or via the search engine on our website here: <https://search.patentstyret.no/>

Patentkrav

1. Forbindelse med formel Ib-iii:

**Ib-iii**

5 eller et farmasøytisk akseptabelt salt derav, hvor:

Ring B er en C6-C10 aryling eller C3-C10 heteroaryl- eller heterocyklisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR;

10 Ring C er en C3-C14 heteroaryl- eller heterocyklisk ring hvor, hvor som helst fra 1 til 4 ringatomer uavhengig er N, O eller S, og hvor ett nitrogen på Ring C er festepunktet til pyridinringen; og hvor, uavhengig for hver forekomst:

15 R₁ er halogen; CN; F₅S; SiR₃; OH; NRR; C1-C6-alkyl eller fluoralkyl; C1-C6-alkoksy eller fluoralkoksy; C1-C6-alkenyl; C1-C6-alkynyl; (C1-C9-alkylen)-R₄ hvor opptil fire CH₂-enheter uavhengig er erstattet med O, CO, S, SO, SO₂ eller NR; C6-C10 aryl; C3-C10 heteroaryl eller heterocyklisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; eller C3-C10 cykloalkyl;

20 R₂ er halogen; OH; NRR; azid; CN; CO₂R; C1-C6-alkyl eller fluoralkyl; C1-C6-alkoksy eller fluoralkoksy; C1-C6-alkenyl; C1-C6-alkynyl; C6-C10 aryl; C3-C13 heteroaryl eller heterocyklisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; C3-C10 cykloalkyl; eller en (C1-C9-alkylen)-R₄ hvor opptil fire CH₂-enheter uavhengig er erstattet med O, CO, S, SO, SO₂ eller NR;

25 eller to R₂-grupper samlet kan danne en =CH₂ eller =O-gruppe;

30 R₃ er halogen; CN; CO₂R; C1-C6-alkyl eller fluoralkyl; C1-C6-alkenyl; C1-C6-alkynyl; C1-C6-alkoksy eller fluoralkoksy; eller C6-C10 aryl; C3-C10 heteroaryl- eller heterocyklisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; C3-C10 cykloalkyl; eller en (C1-C9-alkylen)-R₄ hvor opptil fire CH₂-enheter uavhengig er erstattet med O, CO, S, SO, SO₂ eller NR;

eller to R₃-grupper samlet kan danne en =CH₂ eller =O gruppe;

R₄ er H; azid; CF₃; CHF₂; OR; CCH; CO₂R; OH; C6-C10 aryl, C3-C10 heteroaryl eller heterocykloalkyl hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; C3-C10 cykloalkyl; NRR, NRCOR, CONRR, CN, halo eller SO₂R;

R er uavhengig H; OH; CO₂H; CO₂C1-C6 alkyl; C1-C6 alkyl; C1-C6 alkenyl; C1-C6 alkynyl; C6-C10 aryl; C3-C10 heteroaryl eller heterocykloalkyl hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; eller C3-C10 cykloalkyl;

n er 0, 1, 2 eller 3;

p er 0, 1, 2 eller 3; og

q er 0, 1, 2, 3, 4 eller 5;

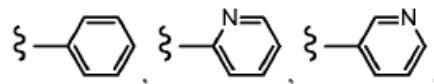
hvor hver av de spesifikke gruppene for variablene R₁-R₄ eventuelt kan være substituert med en eller flere grupper valgt fra halogen, fosfo, OH, cykloalkyl, heterocykloalkyl, aryl, heteroaryl, fluoroalkyl, alkyl, alkenyl, alkynyl, nitro, CN, hydroxyl og (C1-C9alkylen)-E hvor opptil 4 CH₂ enheter uavhengig er erstattet med O, S, SO₂, SO, CO, NH, N-alkyl, N-alkenyl eller N-alkynyl, og E er H, aryl, cycloalkyl, heterocycloalkyl, heteroaryl, alkoxsy, CN eller CF₃, videre hvor hver av aryl, cycloalkyl, heterocycloalkyl og heteroaryl eventuelt er substituert med en eller flere grupper valgt fra halogen, alkyl, amino, CN, alkenyl, alkynyl og alkoxsy; og

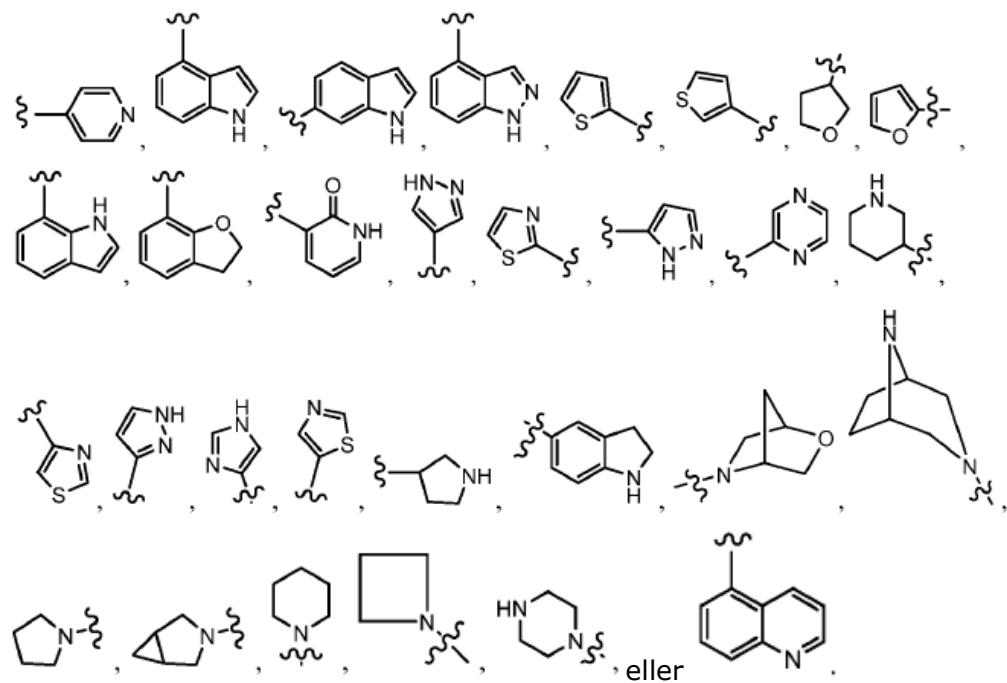
når to alkoxsygrupper er bundet til det samme atom eller tilstøtende atomer, kan de to alkoxsygruppene danne en ring sammen med atomet/atomene de er bundet til; og

hvor uttrykket "amino" refererer til NH₂ som eventuelt er substituert med en eller to grupper uavhengig valgt fra alkyl, cykloalkyl og heterocykloalkyl.

2. Forbindelse eller salt ifølge krav 1, hvor ring B er:

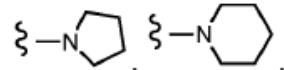
(a) fenyl, pyridyl, pyridin-2(1H)-on, pyrazol, indol, aza-indol, tiofen, dihydrobenzofuran eller kinolin; eller (b) valgt fra



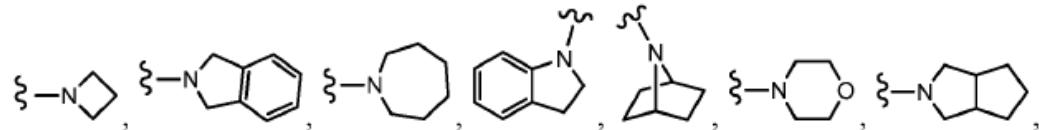


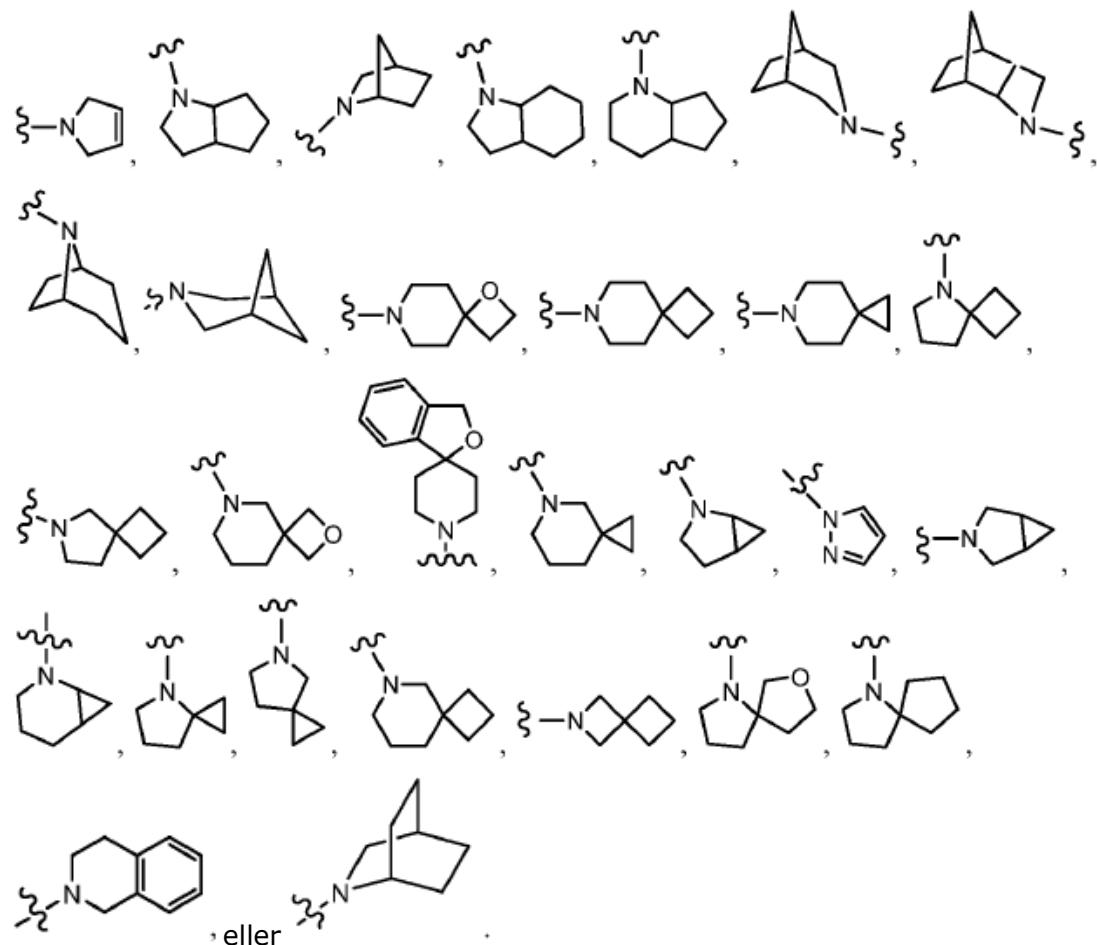
3. Forbindelse eller salt ifølge krav 1 eller krav 2, hvor ring C er valgt fra indol,
5 piperidin, azepan, azetadin, indolin, isoindolin eller pyrrolidin.

4. Forbindelse eller salt ifølge et hvilket som helst av kravene 1 til 3,

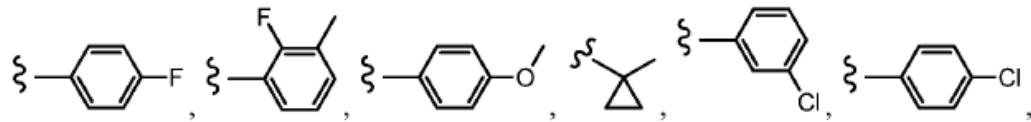
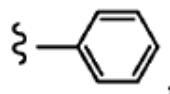


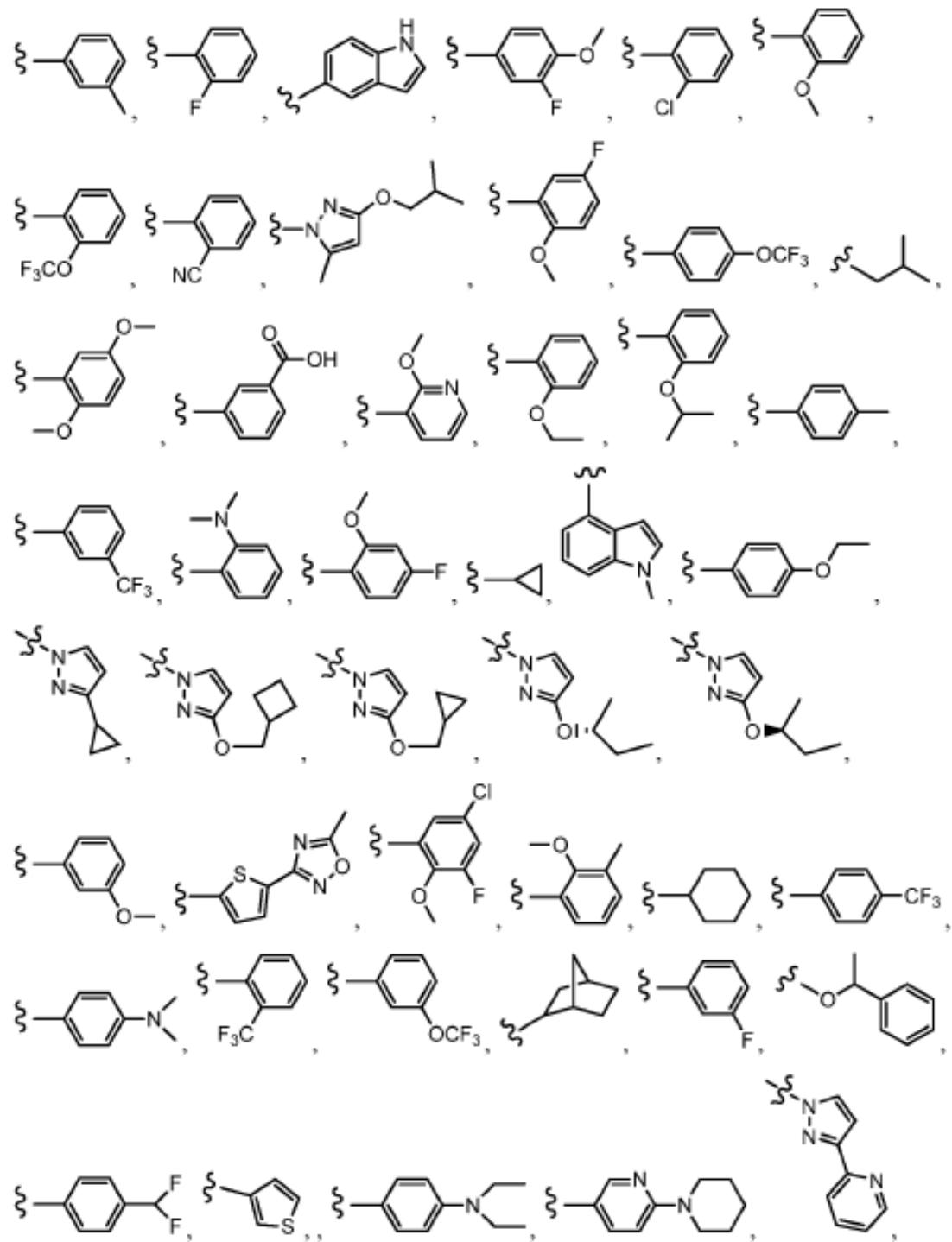
karakterisert ved at ring C er

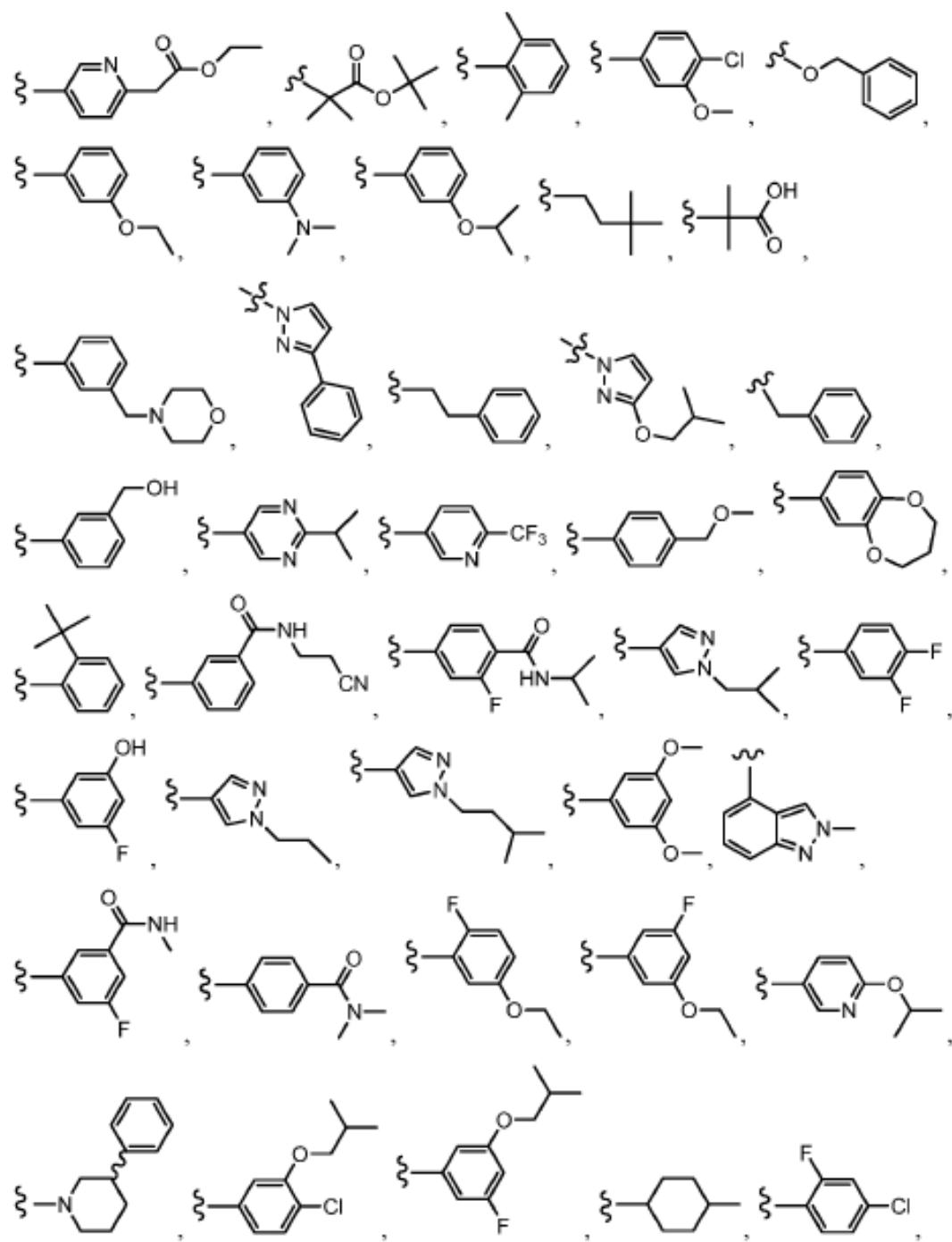


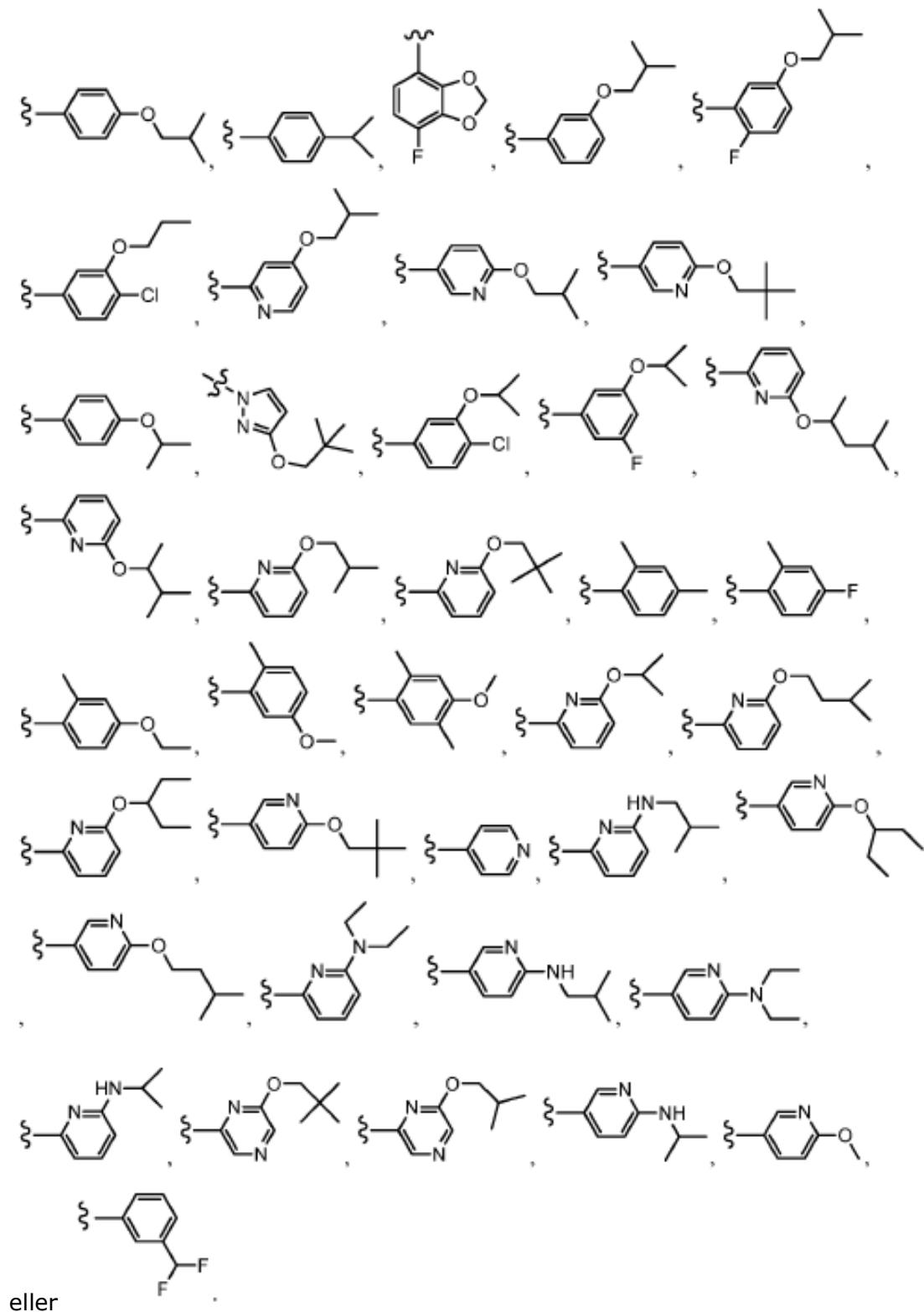


5. Forbindelse eller salt ifølge et hvilket som helst av kravene 1 til 4, hvor R₁
er: (a) halogen, CN, C1-C6-alkyl, C1-C6-alkoksy, C3-C8-cycloalkyl, eller en fenyl,
pyridyl, pyrimidin, indol, aza-indol, pyrazol eller tiofen ring, eller en (C1-C9
alkylen)-R₄ hvor opptil fire CH₂-enheter uavhengig er erstattet med O, CO, S, SO,
SO₂ eller NR, hvor alle ringer kan være substituert med halogen, C1-C6-alkyl, C1-
C6-alkoksy, C1-C6-fluoralkyl, C1-C6-fluoralkoksy, OH, CH₂OH, CH₂OCH₃, CN, CO₂H,
10 amino, amido, C3-C10 heteroaryl eller C3-C10 heterocykloalkyl; eller (b) R₁ er
valgt fra CH₃, Cl, F, CN, OCH₃, CF₃, CH₂CH₃, tBu, CH(CH₃)₂, OCH₂CH₂OCH₂CH₃,







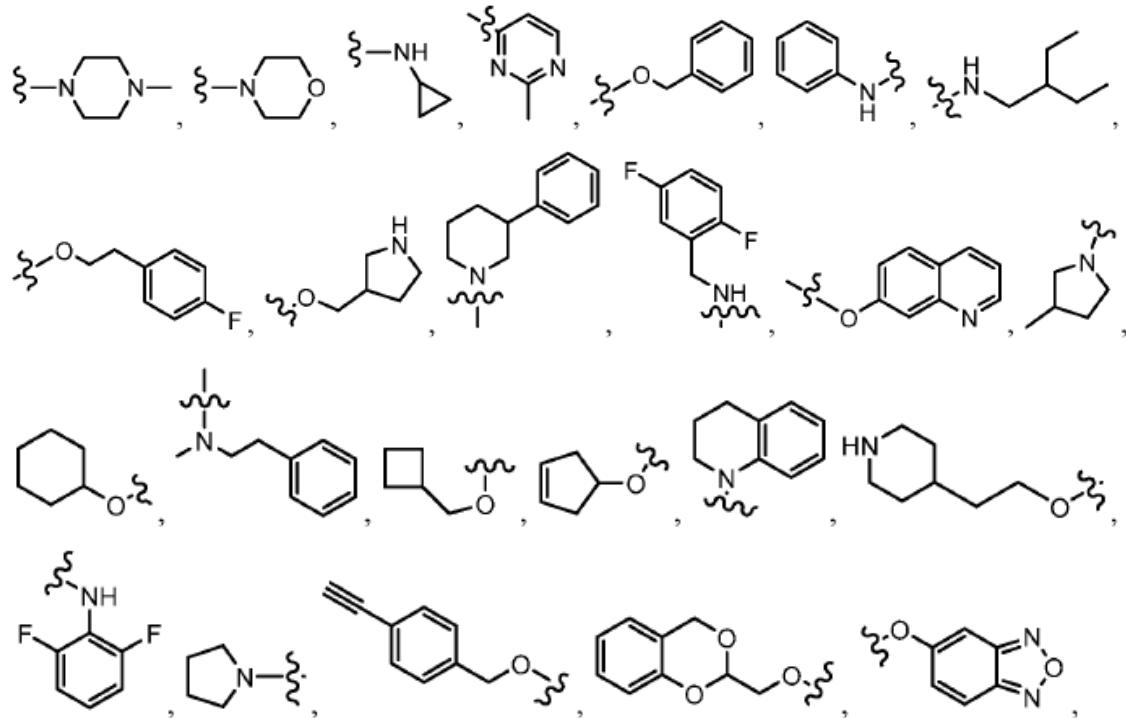


5 6. Forbindelse eller salt ifølge et hvilket som helst av kravene 1 til 5, hvor R₂ er: (a) valgt fra halogen, OH, CN, azid, amino, C1-C6-alkyl eller fluoroalkyl, C1-C6-alkoksy eller fluoroalkoksy, C3-C10-heterocyklig ring hvor opptil 4 ringatomer kan erstattes med O, S, N eller NR; eller en (C1-C9-alkylen)-R₄ hvor opptil fire CH₂-enheter uavhengig er erstattet med O, CO, S, SO, SO₂ eller NR; eller (b) valgt fra

Cl, F, OH, CN, N₃, NH₂, NH(CH₃), N(CH₃)₂, N(CH₃)CH₂CH₂CH₃,
 N(CH₃)CH₂CH₂CH₂CH₃, CH₃, CH₂OH, CH₂CH₃, CH₂CH₂CH₃, =O, CH₃SO₂, CH₃SO₂NH,
 CF₃CONH, CH₃CONH, CH₃CON(CH₃), tBuOCONH, (CH₃)₂CHOCONH, CH(CH₃)₂, CHF₂,

OCH₃, OCH₂CH₃, OCH₂CH₂CH₃, OCH₂CH₂CH(CH₃)₂, OCF₃, OCHF₂, OC(CH₃)₃,

- 5 OCH₂CH₂tBu, NHCH(CH₃)(CH₂CH₂CH₃), OCH(CH₃)₂, NH(CH₂)₂O(CH₂)₂CH₃, C(O)CH₃,
 CH₂CH₂OH, CH₂NH₂, NH(CH₂)₂OH, N(CH₃)CH₂CH₂OCH₃, NHCH₂CH₂COOH,
 NH(CH₂)₂N(CH₃)₂, NH(CH₂)₂NH₂, NH(CH₂)₃NH₂, NH(CH₂)₂OCH₃, NHCH(CH₃)₂,

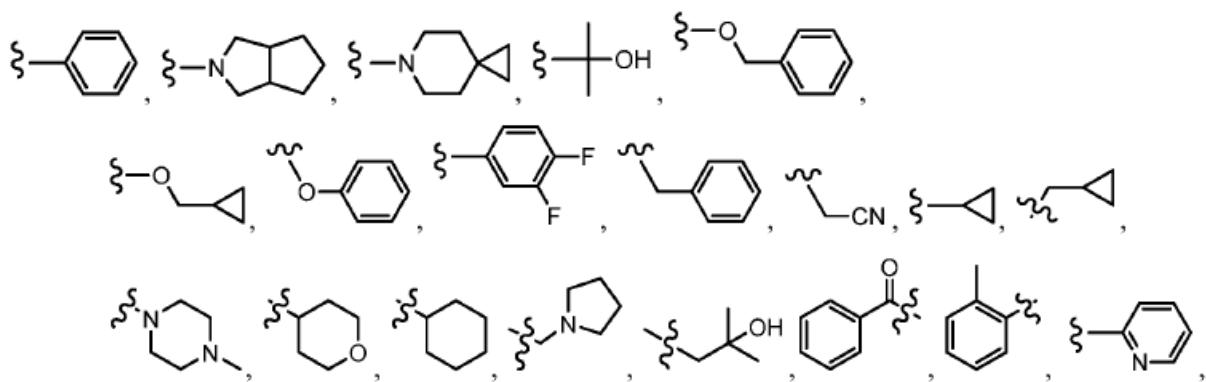


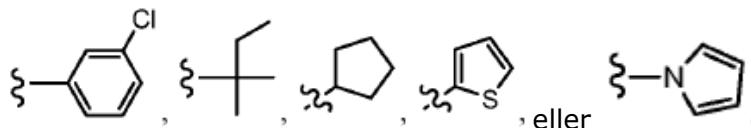
10 eller CO₂H.

7. Forbindelse eller salt ifølge et hvilket som helst av kravene 1 til 6, hvor R₃ er: (a) valgt fra halogen, CN, C1-C6-alkyl eller fluoroalkyl, C1-C6-alkoksy eller C3-

C10 heteroaryl hvor opptil 4 ringatomer kan erstattes av O, S, N eller NR; eller (b)

- 15 valgt fra Cl, I, deuterium, F, CN, CH₃, OH, OCH₃, CF₃, CH₂CH₃, CH₂CF₃, CH₂CH₂CH₃,
 OCH₂CH(CH₃)₂, OCH(CH₃)₂, CO₂H, CO₂NH₂, OCH₂CH₃, CH₂OCH₃, CH(CH₃)₂, CCH,
 CH₂CONH₂, CO₂CH₃, -CH₂N(CH₃)₂, CO₂tBu, tBu, =CH₂, =O,





8. Forbindelse eller salt ifølge et hvilket som helst av kravene 1 til 7, hvor n er:

- (a) 0; eller
- (b) 1; eller
- (c) 2.

9. Forbindelse eller salt ifølge et hvilket som helst av kravene 1 til 8, hvor p er:

- (a) 0; eller
- (b) 1; eller
- (c) 2.

10. Forbindelse eller salt ifølge krav 1, hvor R_1 er fenyl, pyridin eller pyrazol, og n er 1.

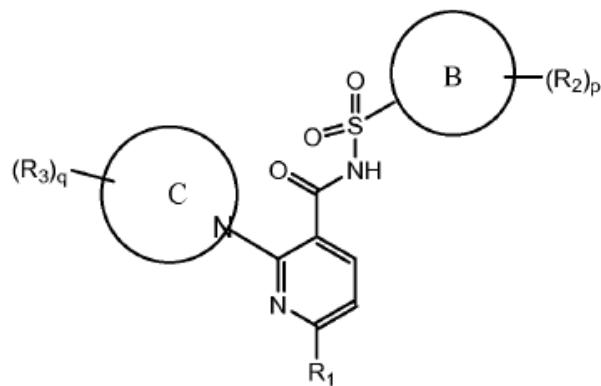
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11. Forbindelse eller salt ifølge krav 1, hvor R_1 er fenyl, pyridin eller pyrazol, n er 1, R_2 er amino eller alkyl, og p er 0 eller 1.

12. Forbindelse eller salt ifølge krav 1, hvor R_1 er fenyl, pyridin eller pyrazol, n er 1, R_3 er alkyl, og q er 1, 2, 3 eller 4.

20

13. Forbindelse ifølge krav 1, hvor forbindelsen er en forbindelse med formel Ib-iv:



25

Ib-iv

eller et farmasøytisk akseptabelt salt derav, hvor uavhengig av hver forekomst:

Ring B er en C6-C10 arylring eller C3-C10 heteroaryl eller heterocyklig ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR;

Ring C er en C3-C14 heteroaryl- eller heterocyklistisk ring hvor, hvor som helst fra 1 til 4 ringatomer uavhengig er N, O eller S, og hvor ett nitrogen på Ring C er festepunktet til pyridinringen;

R₁ er C6-C10 aryl eller C3-C10 heteroaryl hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR;

R₂ er halogen; OH; NRR; azid; CN; CO₂R; C1-C6-alkyl eller fluoralkyl; C1-C6-alkoksy eller fluoralkoksy; C1-C6-alkenyl; C1-C6-alkynyl; C6-C10 aryl; C3-C13 heteroaryl eller heterocyklistisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; C3-C10 cykloalkyl; eller en (C1-C9-alkylen)-R₄ hvor opptil fire CH₂-enheter uavhengig er erstattet med O, CO, S, SO, SO₂ eller NR;

eller to R₂-grupper samlet kan danne en =CH₂ eller =O-gruppe;

R₃ er halogen; CN; CO₂R; C1-C6-alkyl eller fluoralkyl; C1-C6-alkenyl; C1-C6-alkynyl; C1-C6-alkoksy eller fluoralkoksy; eller C6-C10 aryl; C3-C10 heteroaryl eller heterocyklistisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; C3-C10 cykloalkyl; eller en (C1-C9-alkylen)-R₄ hvor opptil fire CH₂-enheter uavhengig er erstattet med O, CO, S, SO, SO₂ eller NR;

eller to R₃-grupper samlet kan danne en =CH₂ eller =O gruppe;

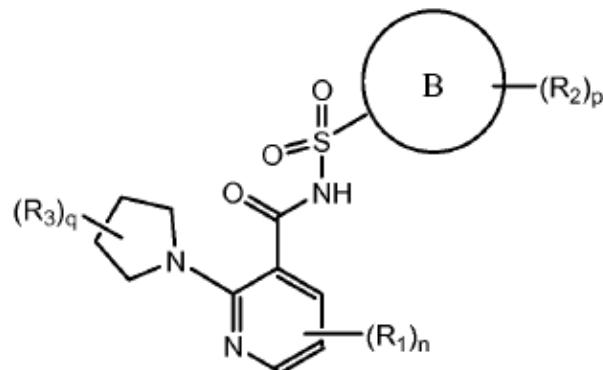
R₄ er H; azid; CF₃; CHF₂; OR; CCH; CO₂R; OH; C6-C10 aryl, C3-C10 heteroaryl eller heterocykloalkyl hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; C3-C10 cykloalkyl; NRR, NRCOR, CONRR, CN, halo eller SO₂R;

R er uavhengig H; OH; CO₂H; CO₂C1-C6 alkyl; C1-C6 alkyl; C1-C6 alkenyl; C1-C6 alkynyl; C6-C10 aryl; C3-C10 heteroaryl eller heterocykloalkyl hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; eller C3-C10 cykloalkyl;

p er 0, 1, 2 eller 3; og

q er 0, 1, 2, 3, 4 eller 5.

14. Forbindelse ifølge krav 1, hvor forbindelsen er en forbindelse med formel Ib-30 v:

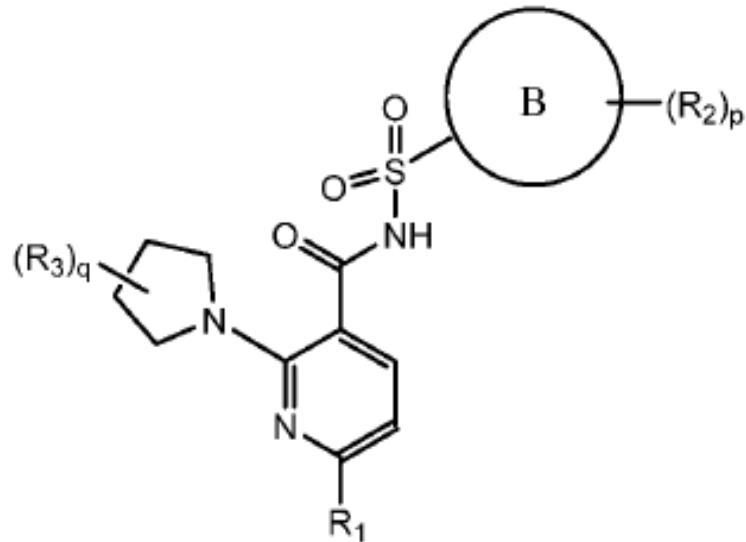
**Ib-v**

eller et farmasøyttisk akseptabelt salt derav, hvor uavhengig av hver forekomst:

- 5 Ring B er en C6-C10 arylring eller C3-C10 heteroaryl eller heterocyklisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR;
- R₁ er halogen; CN; F₅S; SiR₃; OH; NRR; C1-C6-alkyl eller fluoralkyl; C1-C6-alkoksy eller fluoralkoksy; C1-C6-alkenyl; C1-C6-alkynyl; (C1-C9-alkylen)-R₄ hvor opptil fire CH₂-enheter uavhengig er erstattet med O, CO, S, SO, SO₂ eller NR; C6-10 C10 aryl; C3-C10 heteroaryl eller heterocyklisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; eller C3-C10 cykloalkyl;
- R₂ er halogen; OH; NRR; azid; CN; CO₂R; C1-C6-alkyl eller fluoralkyl; C1-C6-alkoksy eller fluoralkoksy; C1-C6-alkenyl; C1-C6-alkynyl; C6-C10 aryl; C3-C13 heteroaryl eller heterocyklisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; C3-C10 cykloalkyl; eller en (C1-C9-alkylen)-R₄ hvor opptil fire CH₂-enheter uavhengig er erstattet med O, CO, S, SO, SO₂ eller NR;
- 15 eller to R₂-grupper samlet kan danne en =CH₂ eller =O gruppe;
- R₃ er halogen; CN; CO₂R; C1-C6-alkyl eller fluoralkyl; C1-C6-alkenyl; C1-C6-alkynyl; C1-C6-alkoksy eller fluoralkoksy; eller C6-C10 aryl; C3-C10 heteroaryl-20 eller heterocyklisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; C3-C10 cykloalkyl; eller en (C1-C9-alkylen)-R₄ hvor opptil fire CH₂-enheter uavhengig er erstattet med O, CO, S, SO, SO₂ eller NR;
- eller to R₃-grupper samlet kan danne en =CH₂ eller =O gruppe;
- R₄ er H; azid; CF₃; CHF₂; OR; CCH; CO₂R; OH; C6-C10 aryl, C3-C10 heteroaryl eller heterocykloalkyl hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; C3-C10 cykloalkyl; NRR, NRCOR, CONRR, CN, halo eller SO₂R;
- 25 R er uavhengig H; OH; CO₂H; CO₂C1-C6 alkyl; C1-C6 alkyl; C1-C6 alkenyl; C1-C6 alkynyl; C6-C10 aryl; C3-C10 heteroaryl eller heterocykloalkyl hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; eller C3-C10 cykloalkyl;

n er 0, 1, 2 eller 3;
 p er 0, 1, 2 eller 3; og
 q er 0, 1, 2, 3, 4 eller 5.

- 5 15. Forbindelse ifølge krav 1, hvor forbindelsen er en forbindelse med formel Ib-vi:

**Ib-vi**

- 10 eller et farmasøytisk akseptabelt salt derav, hvor uavhengig av hver forekomst:

Ring B er en C6-C10 arylring eller C3-C10 heteroaryl eller heterocyklisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR;

- 15 R₁ er C6-C10 aryl eller C3-C10 heteroaryl hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR;

R₂ er halogen; OH; NRR; azid; CN; CO₂R; C1-C6-alkyl eller fluoralkyl; C1-C6-alkoksy eller fluoralkoksy; C1-C6-alkenyl; C1-C6-alkynyl; C6-C10 aryl; C3-C13 heteroaryl eller heterocyklisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; C3-C10 cykloalkyl; eller en (C1-C9-alkylen)-R₄ hvor opptil fire CH₂-enheter uavhengig er erstattet med O, CO, S, SO, SO₂ eller NR;

20 eller to R₂-grupper samlet kan danne en =CH₂ eller =O-gruppe;

R₃ er halogen; CN; CO₂R; C1-C6-alkyl eller fluoralkyl; C1-C6-alkenyl; C1-C6-alkynyl; C1-C6-alkoksy eller fluoralkoksy; eller C6-C10 aryl; C3-C10 heteroaryl- eller heterocyklisk ring hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; C3-C10 cykloalkyl; eller en (C1-C9-alkylen)-R₄ hvor opptil fire CH₂-enheter uavhengig er erstattet med O, CO, S, SO, SO₂ eller NR;

eller to R₃-grupper samlet kan danne en =CH₂ eller =O gruppe;

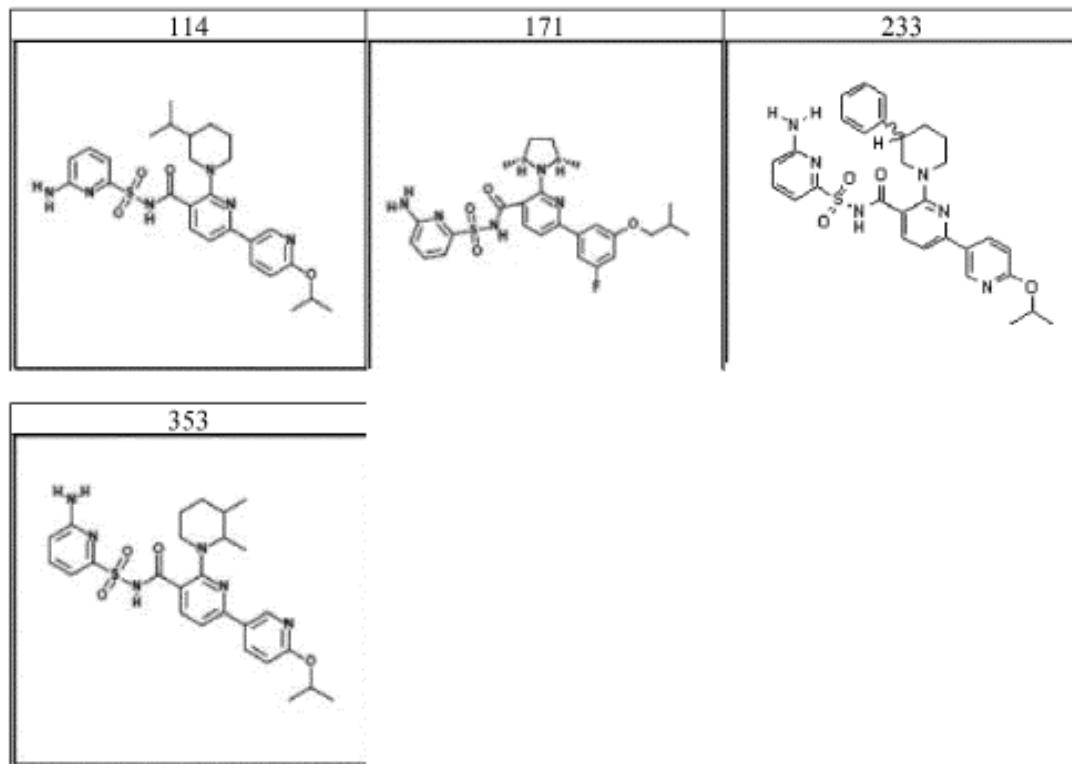
R_4 er H; azid; CF_3 ; CHF_2 ; OR; CCH; CO_2R ; OH; C6-C10 aryl, C3-C10 heteroaryl eller heterocykloalkyl hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; C3-C10 cykloalkyl; NRR, NRCOR, CONRR, CN, halo eller SO_2R ;

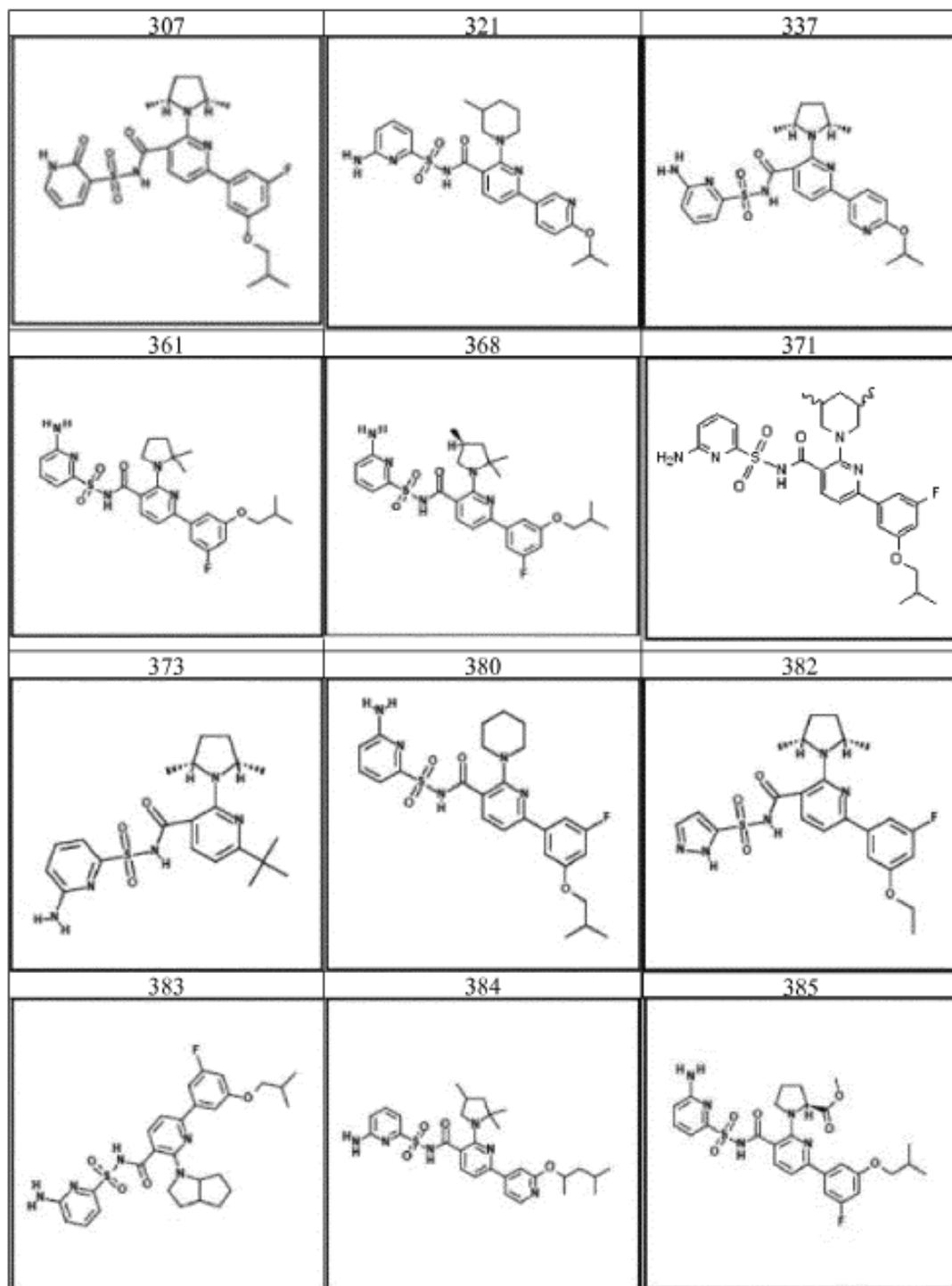
5 R er uavhengig H; OH; CO_2H ; $CO_2C_1-C_6$ alkyl; C1-C6 alkyl; C1-C6 alkenyl; C1-C6 alkynyl; C6-C10 aryl; C3-C10 heteroaryl eller heterocykloalkyl hvor hvor som helst fra 1 til 4 ringatomer uavhengig er O, S, N eller NR; eller C3-C10 cykloalkyl;

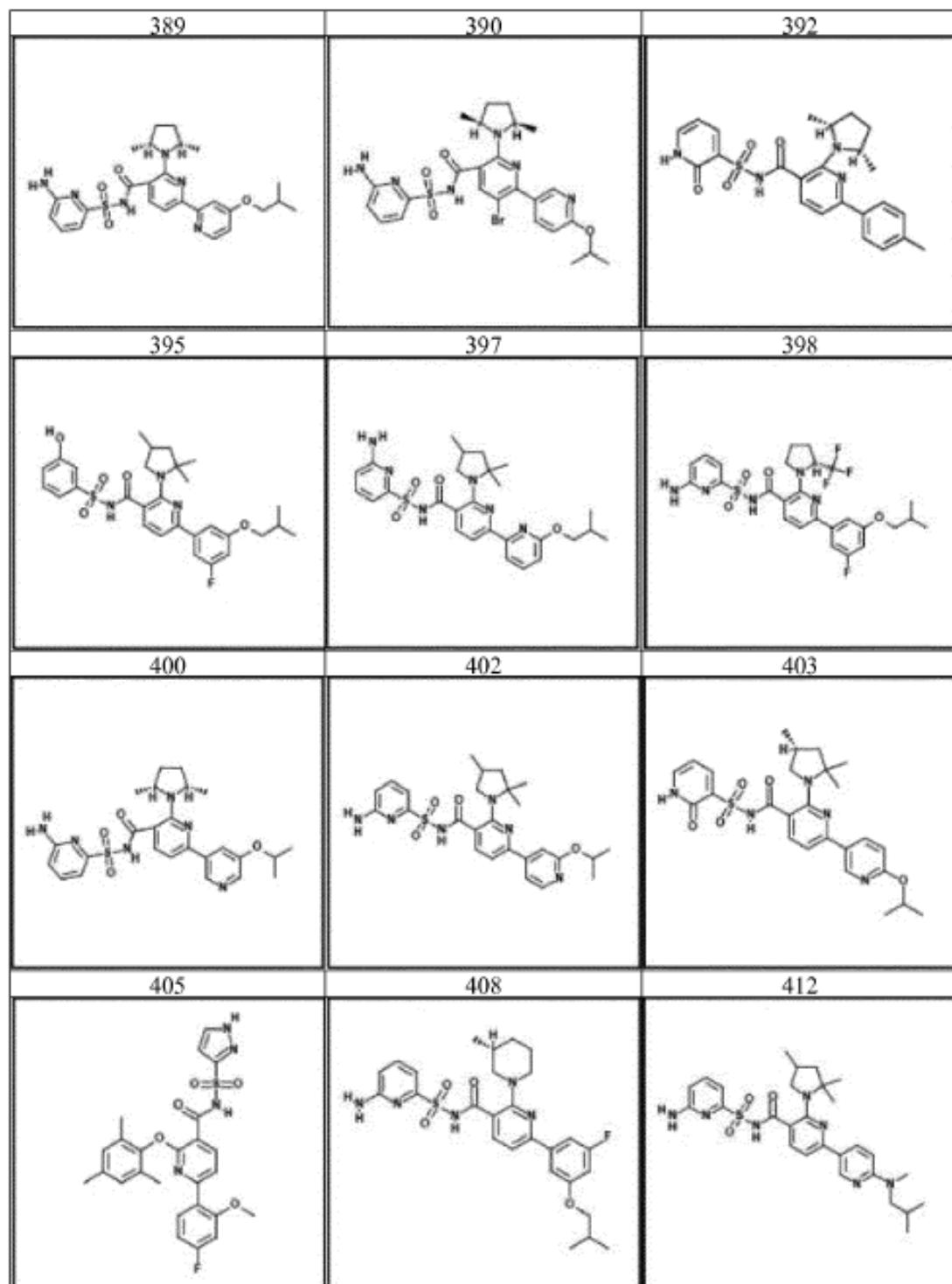
 p er 0, 1, 2 eller 3; og

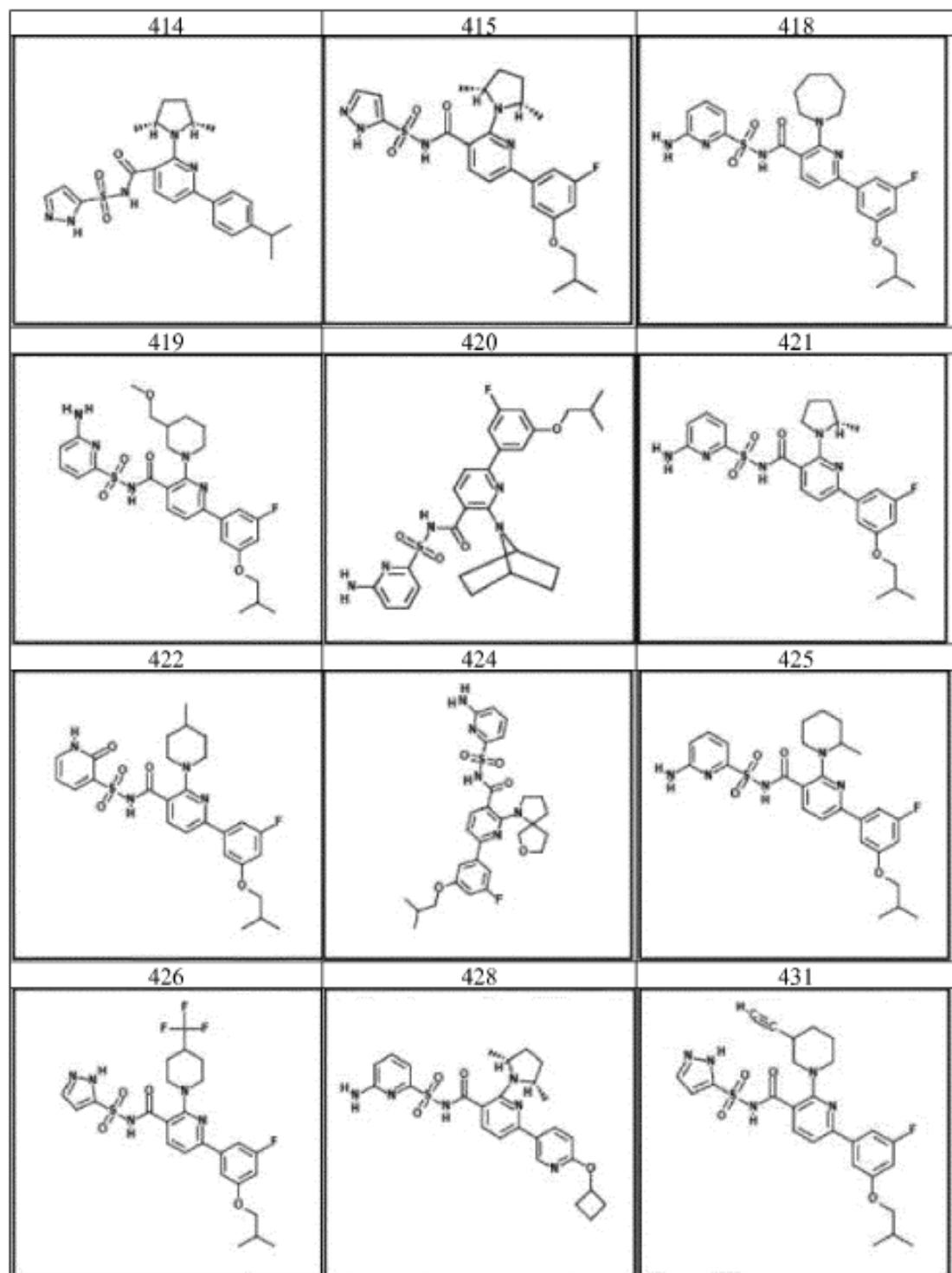
10 q er 0, 1, 2, 3, 4 eller 5.

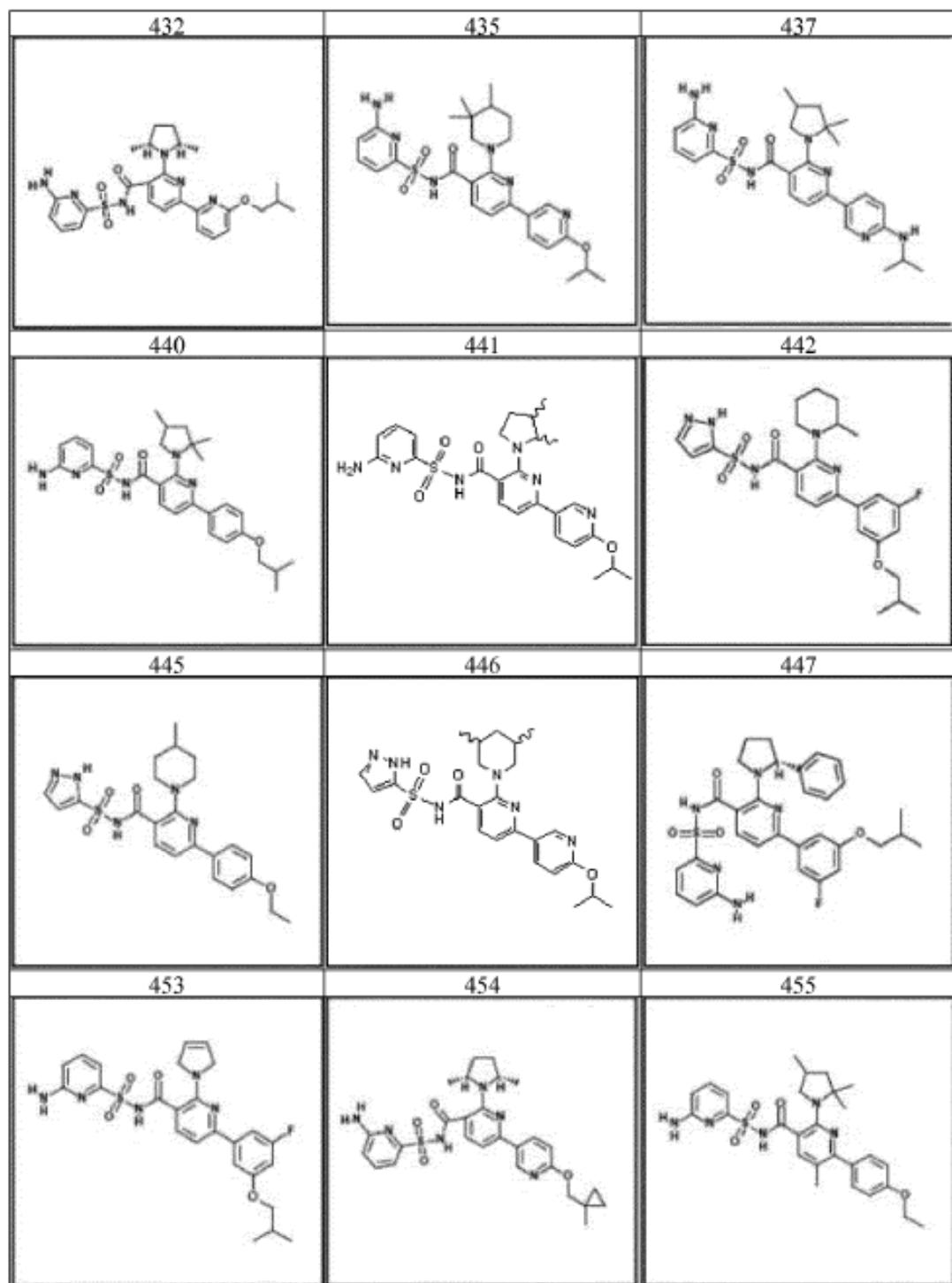
16. Forbindelse ifølge krav 1, eller et farmasøytisk akseptabelt salt derav, hvor forbindelsen er valgt fra:

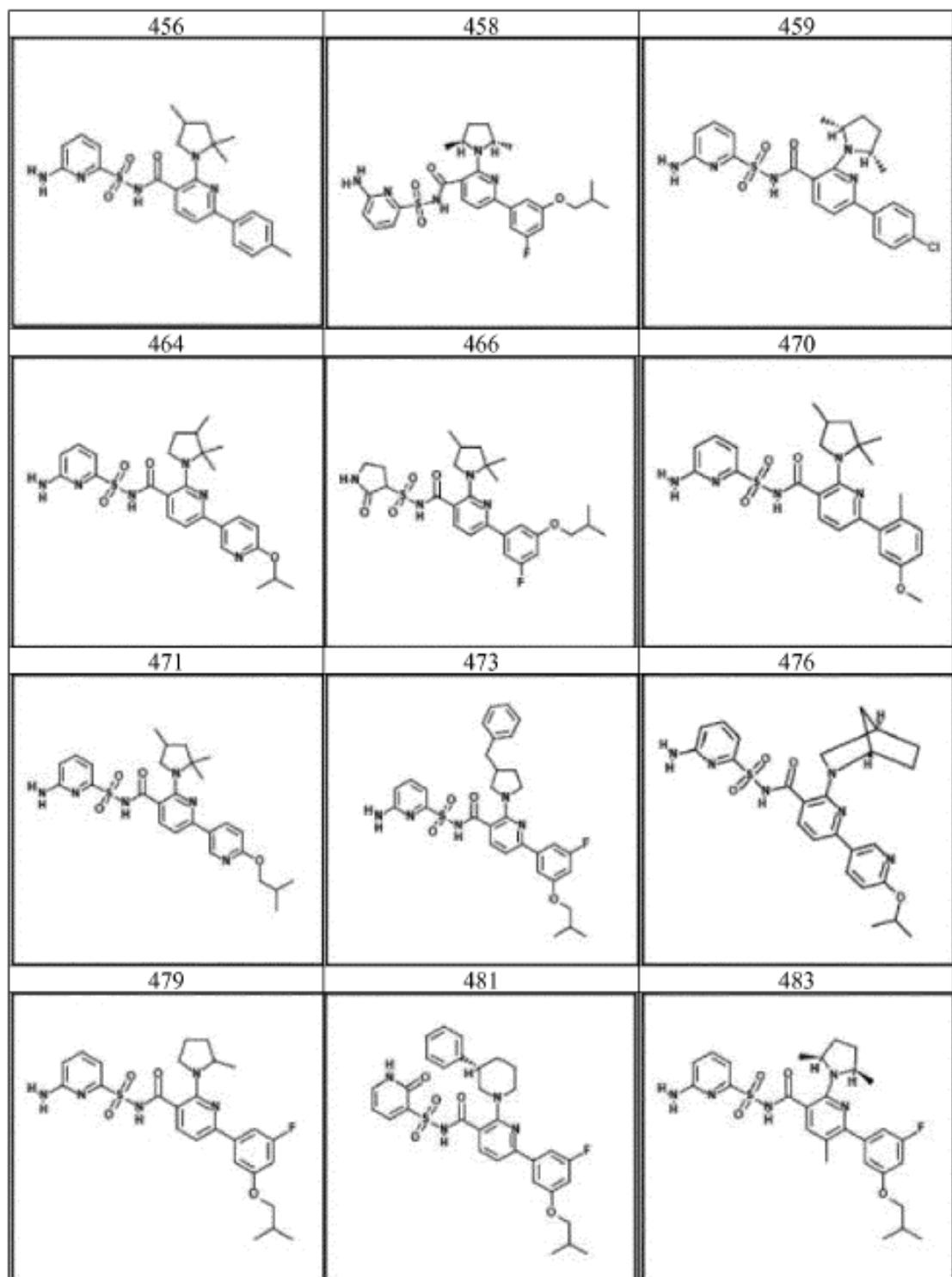


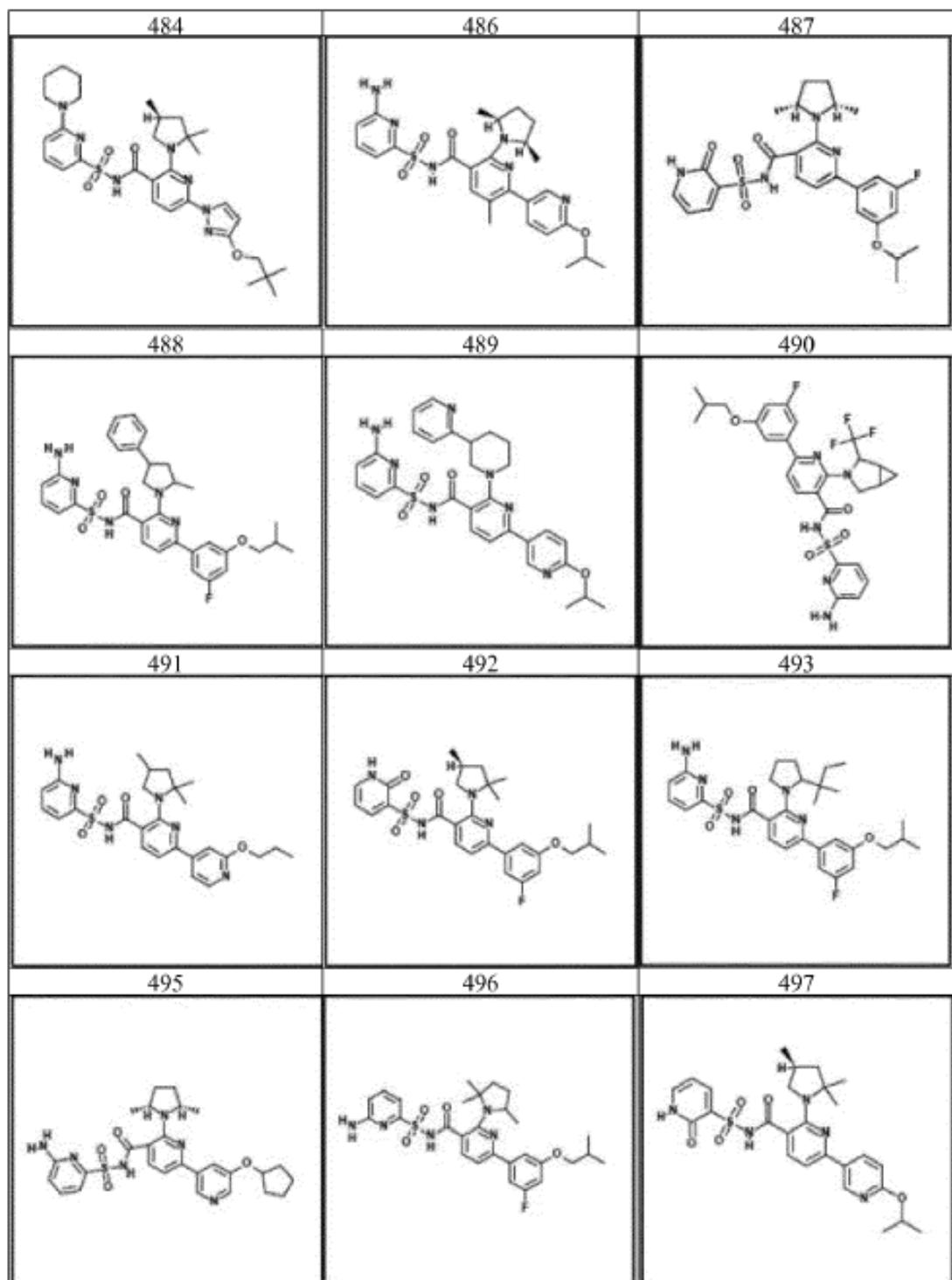


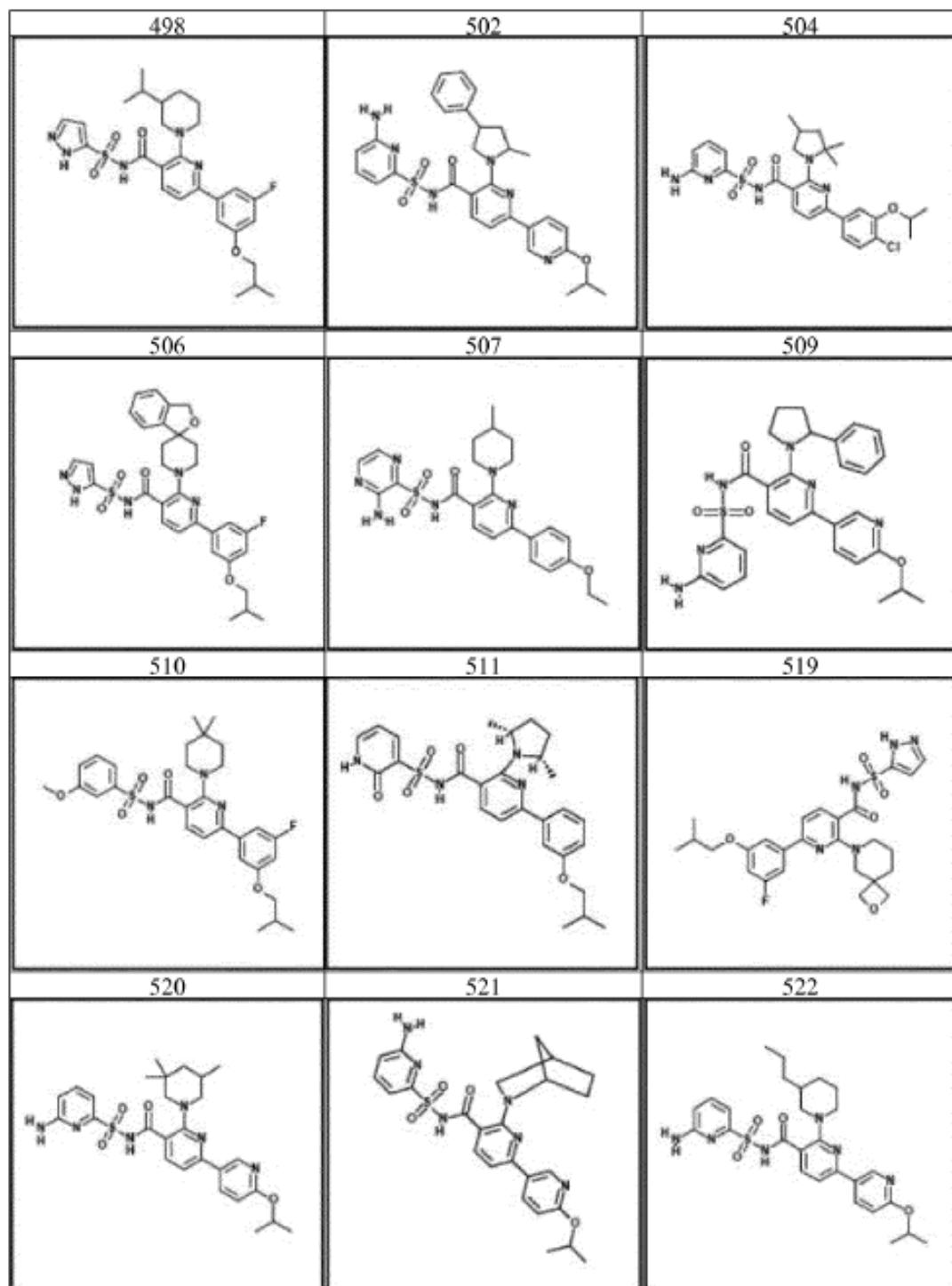


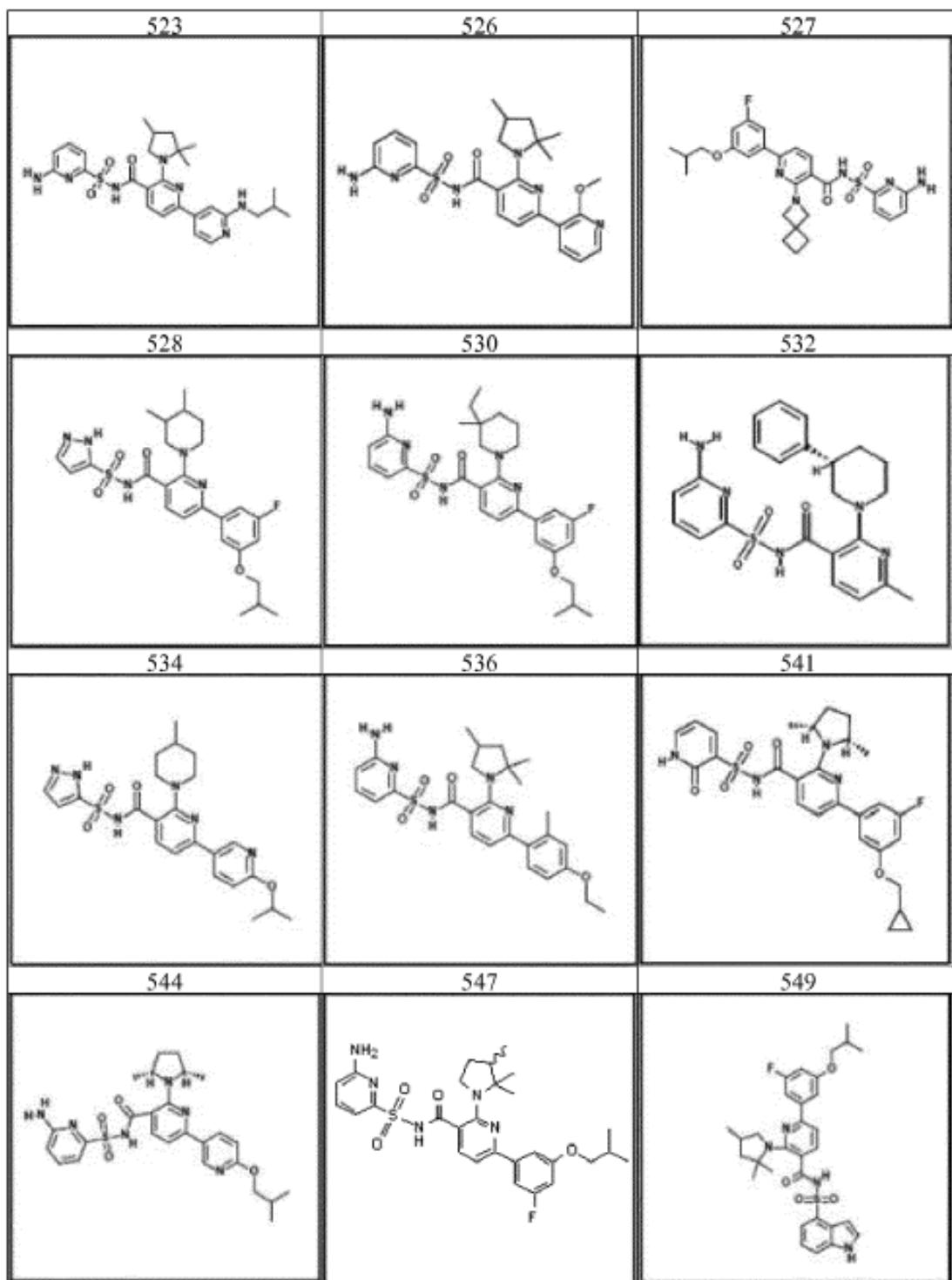


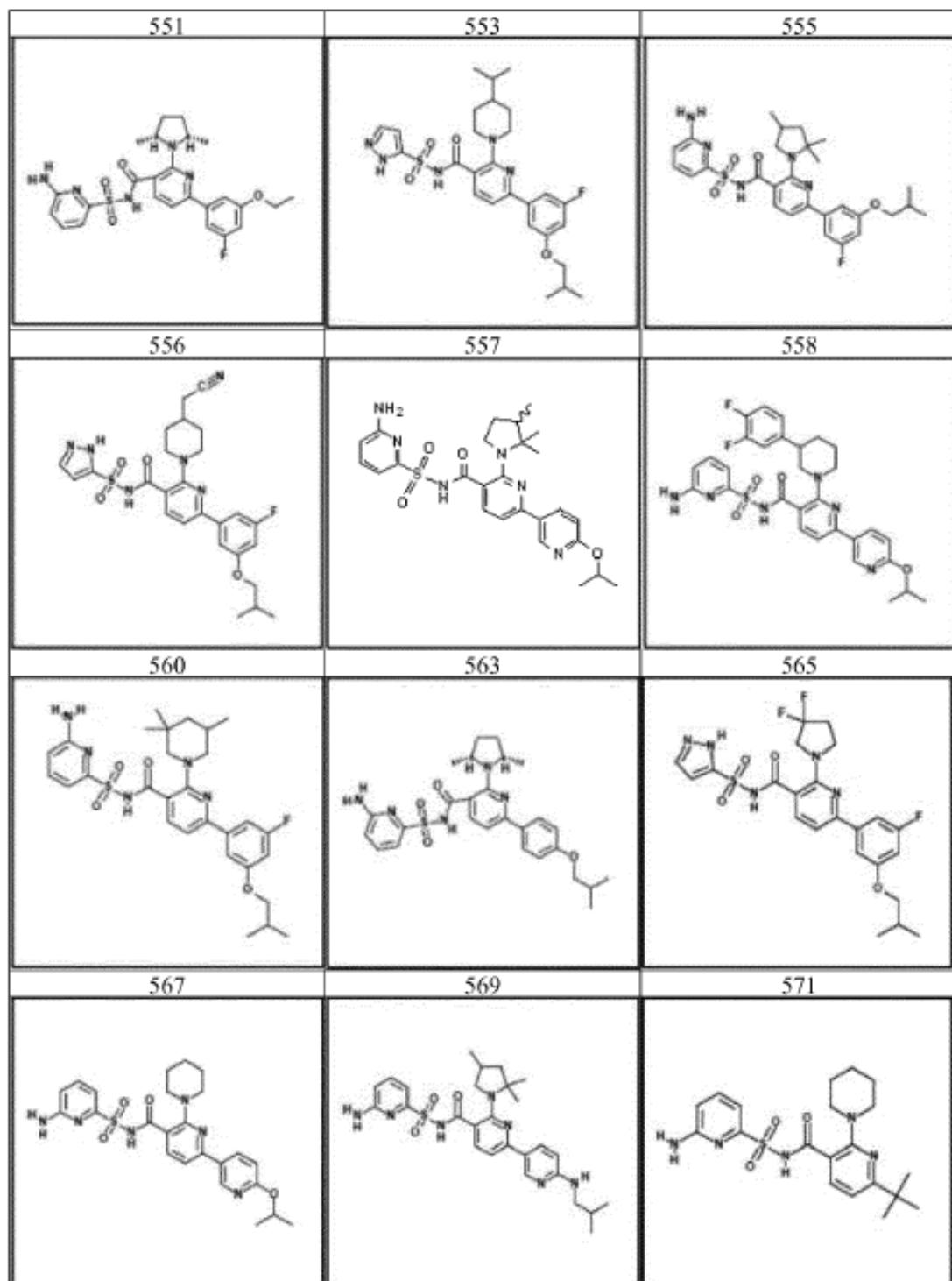


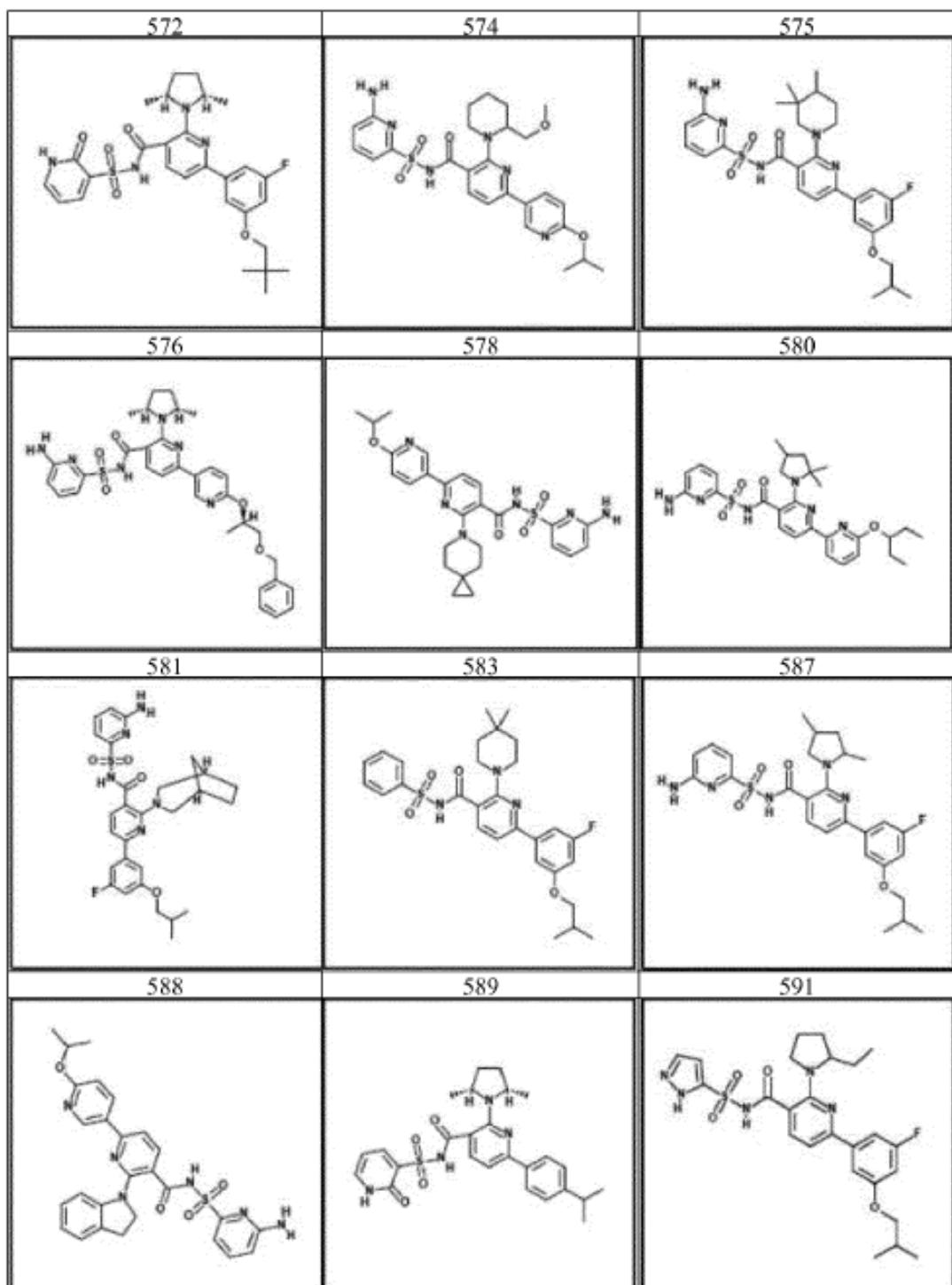


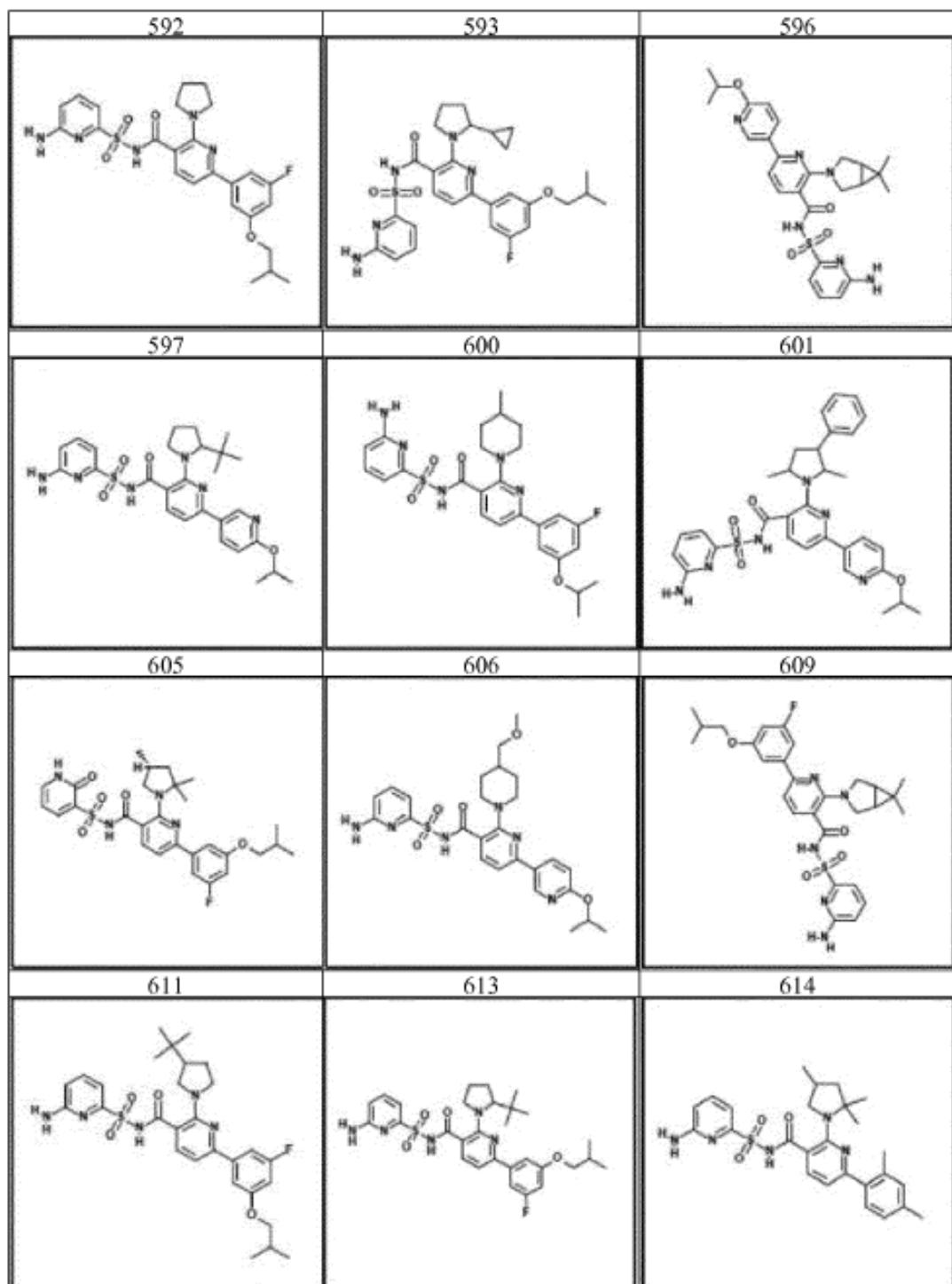


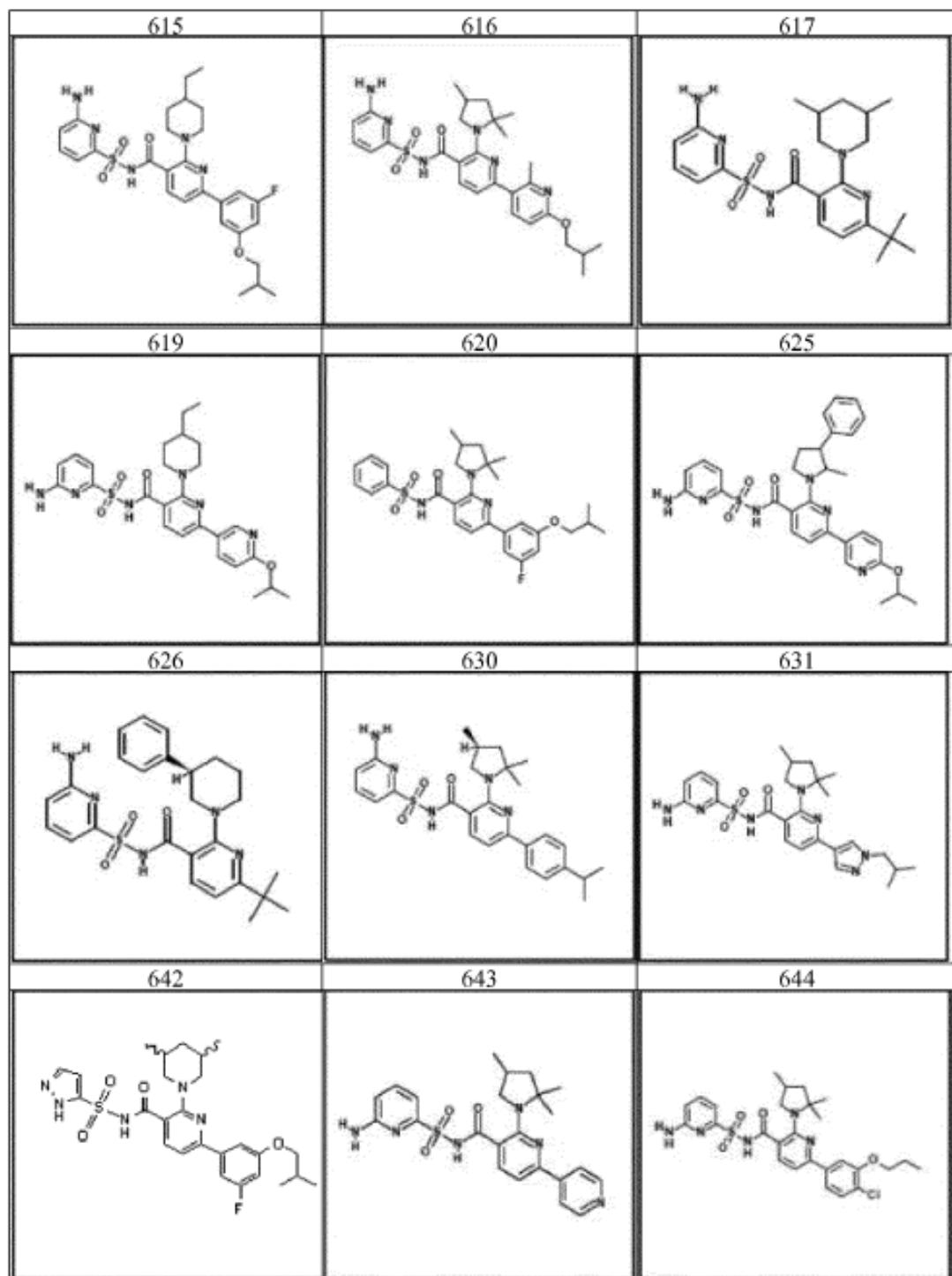


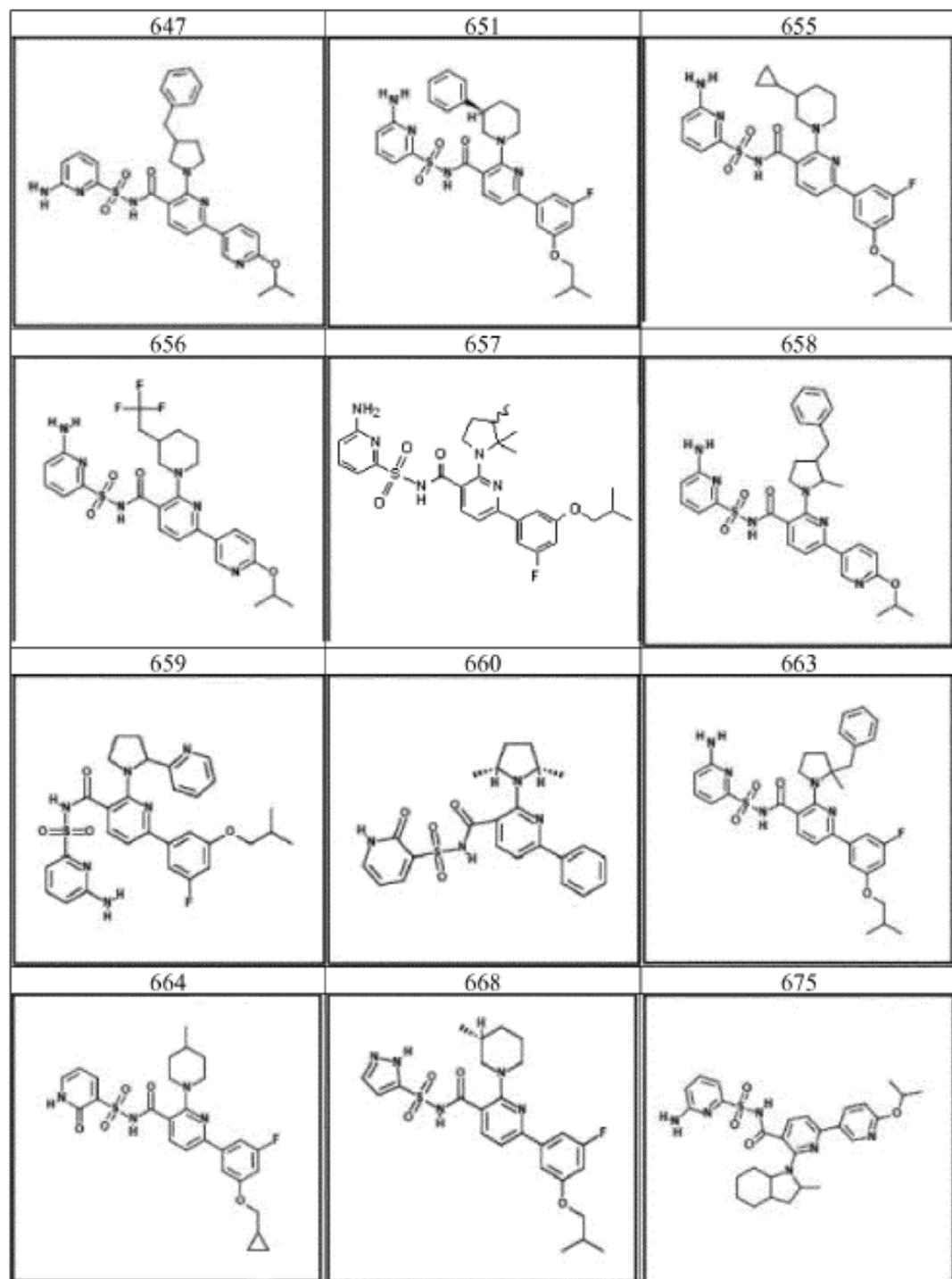


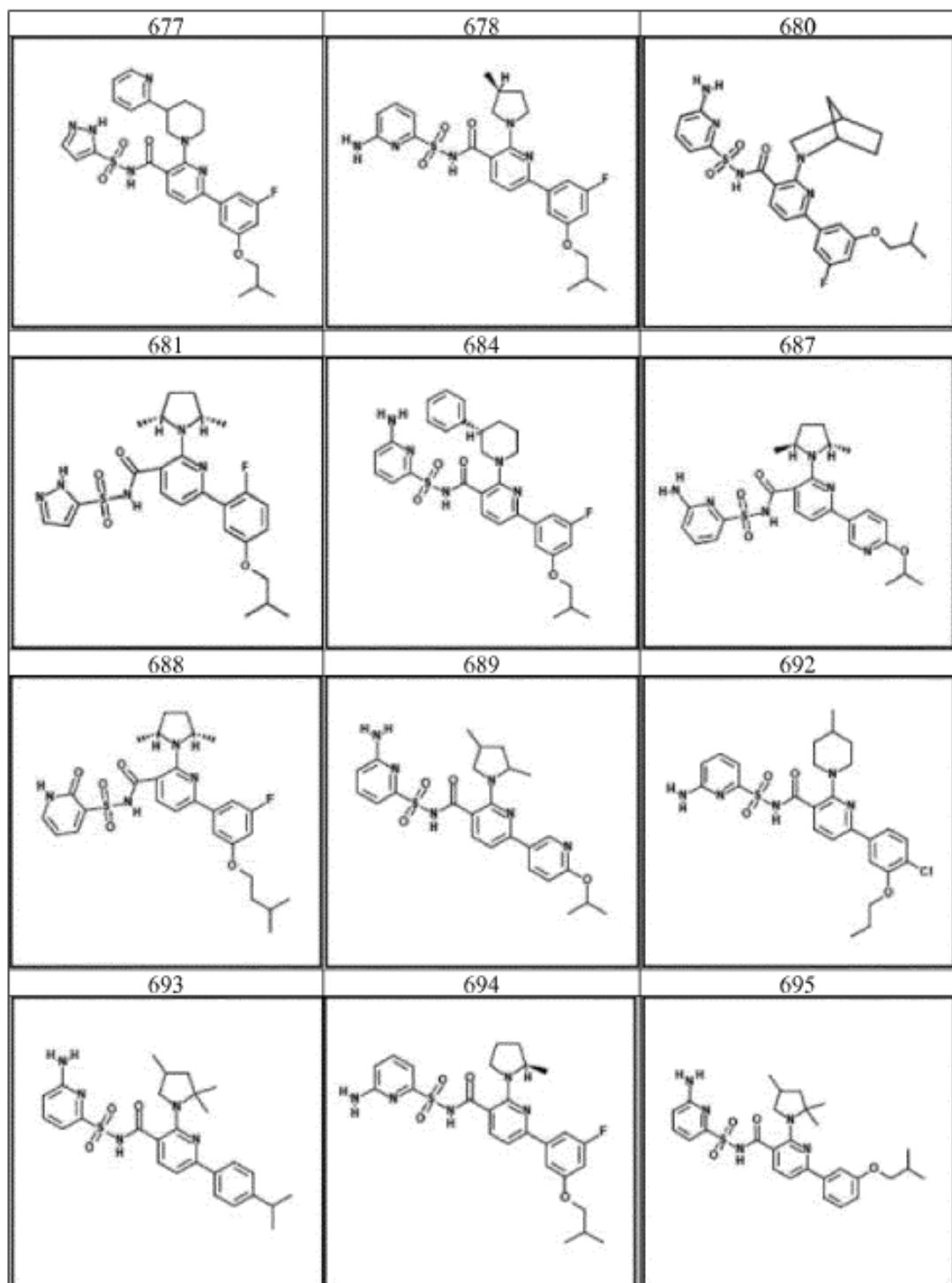


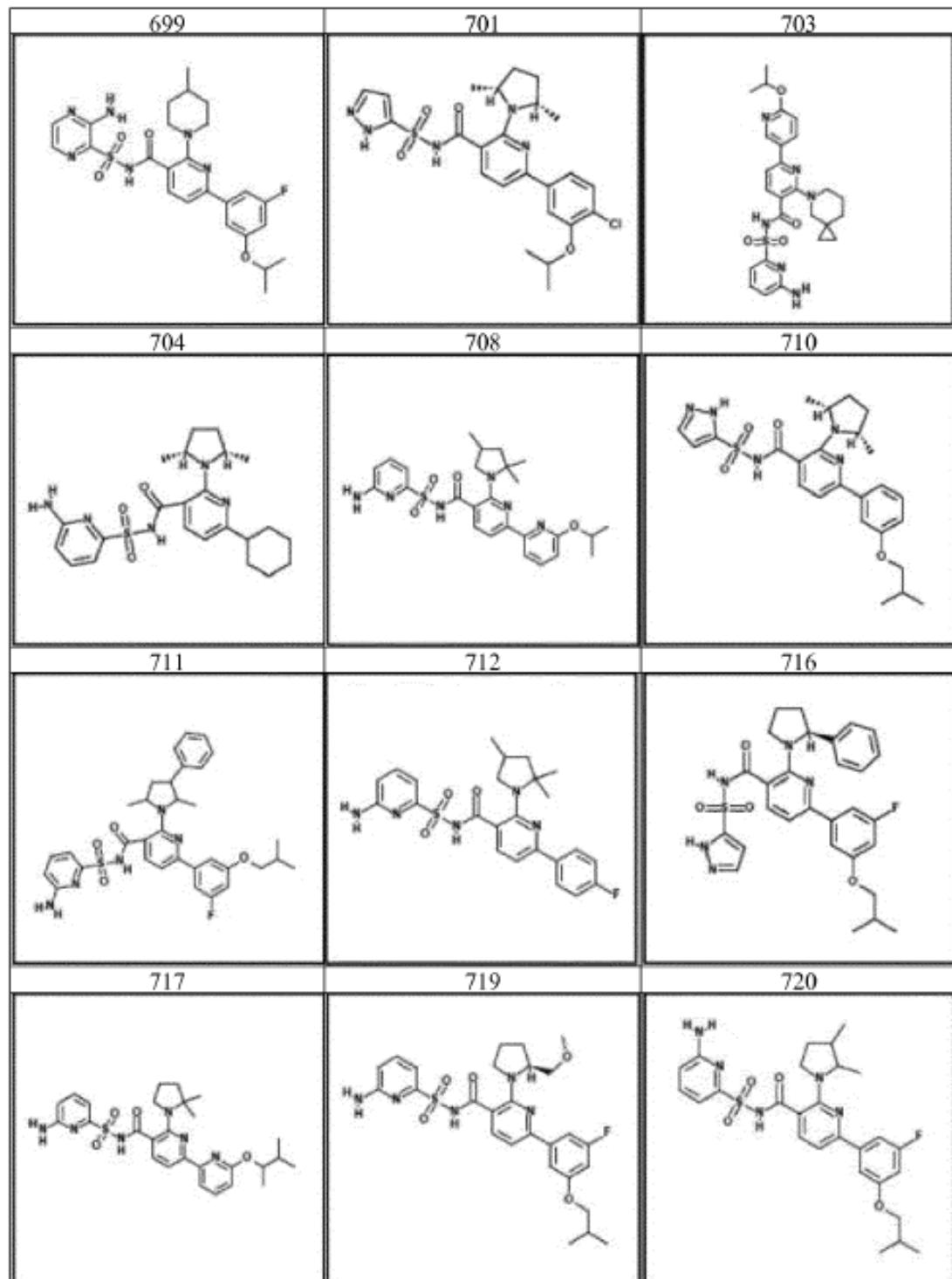


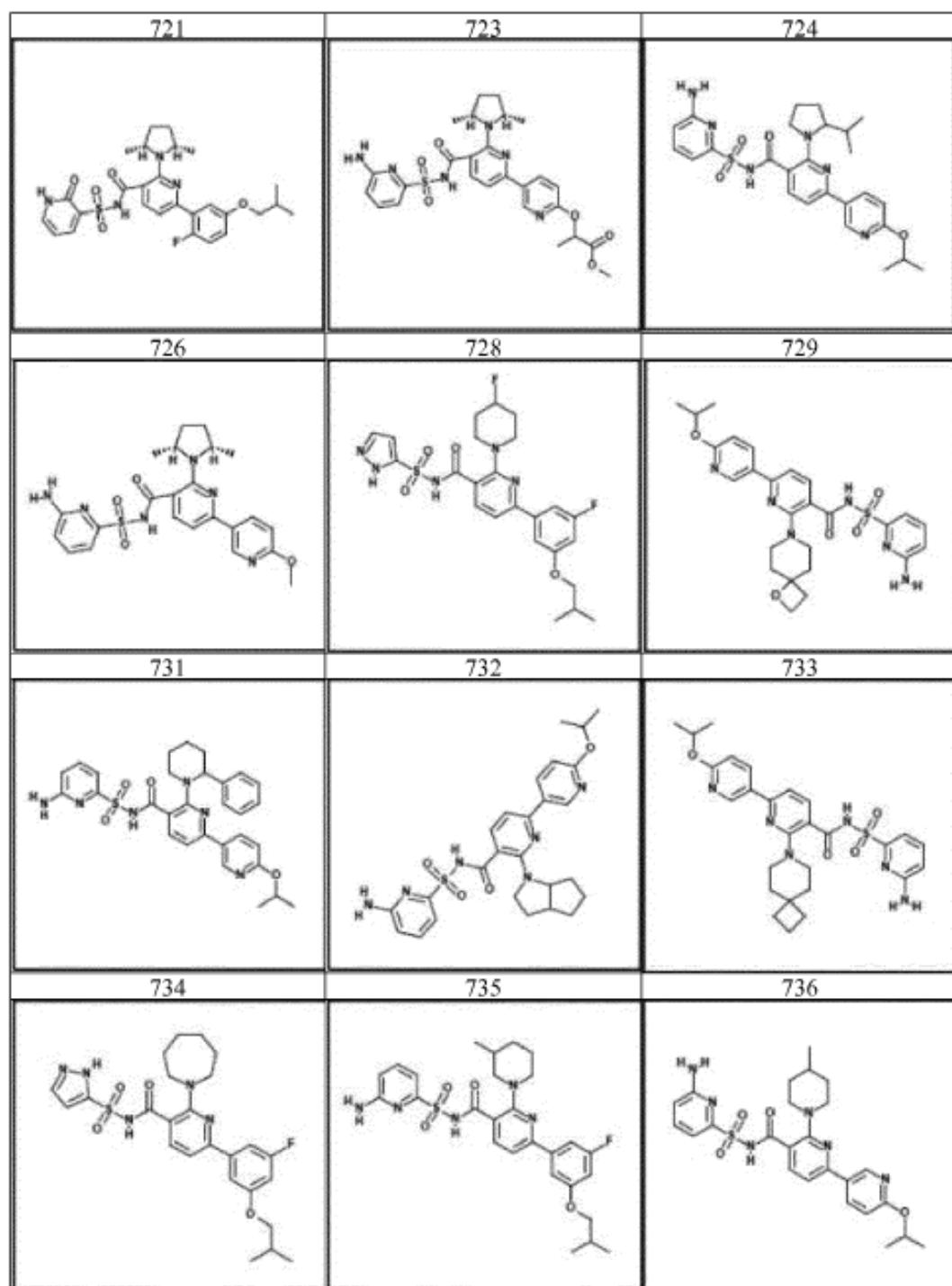


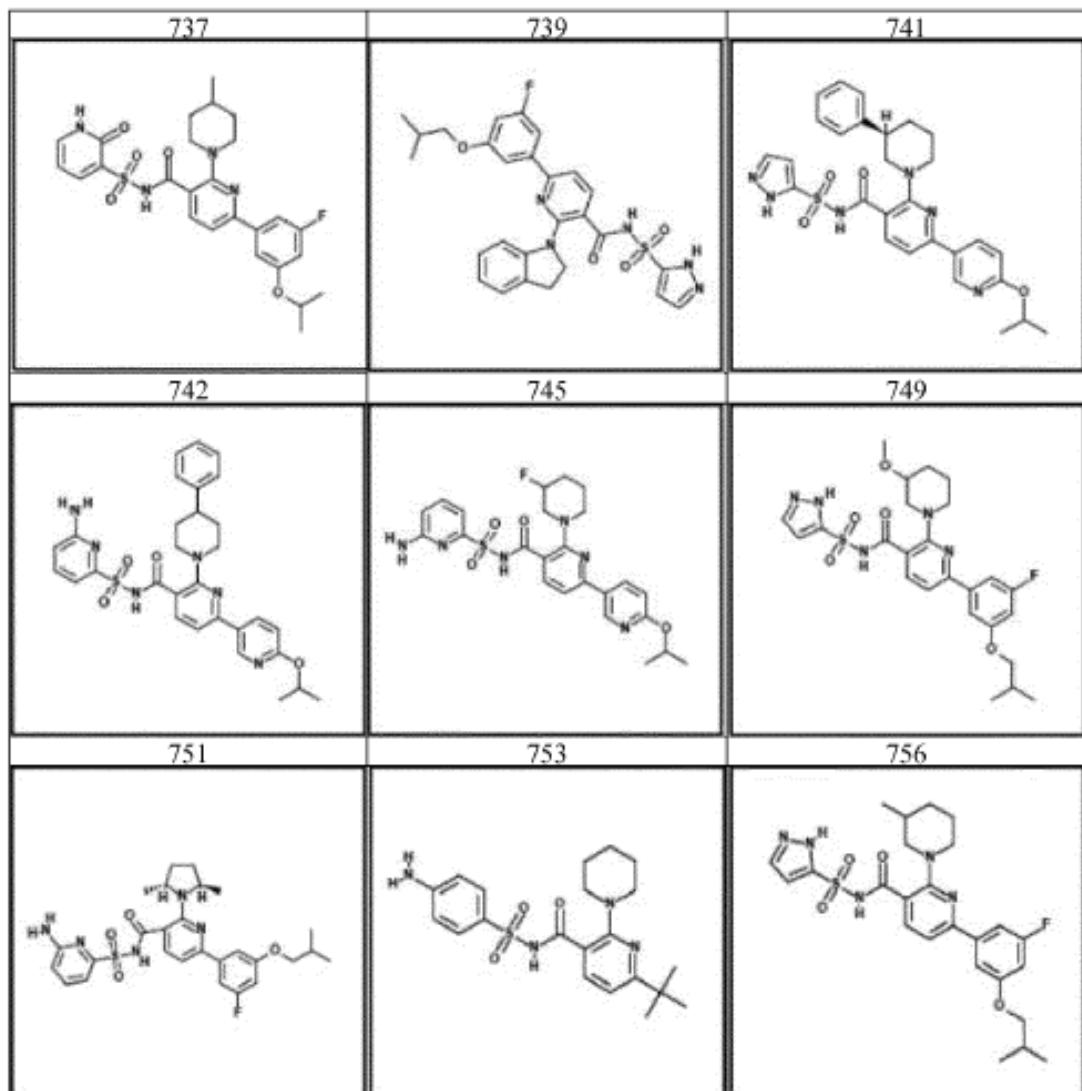


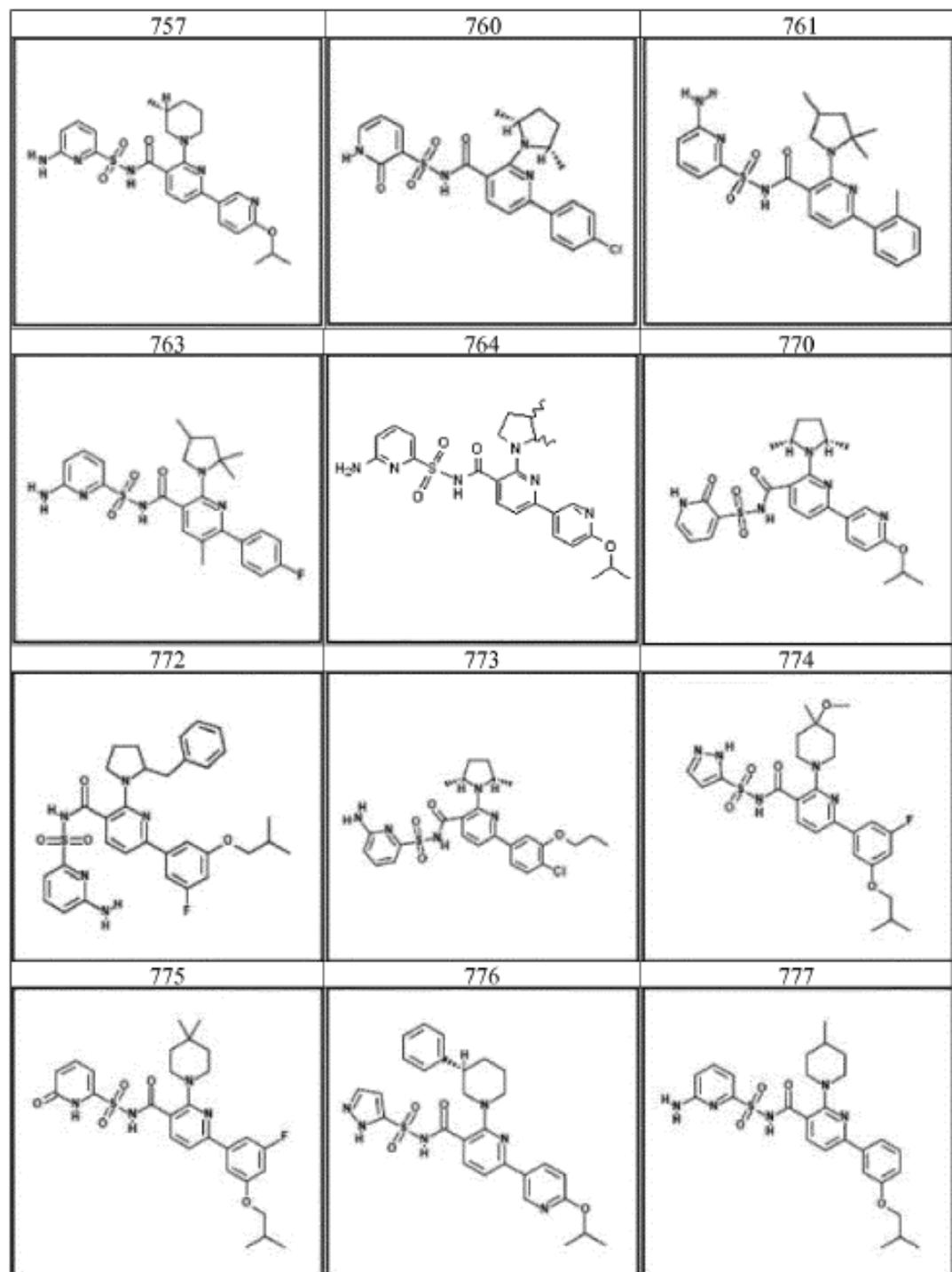


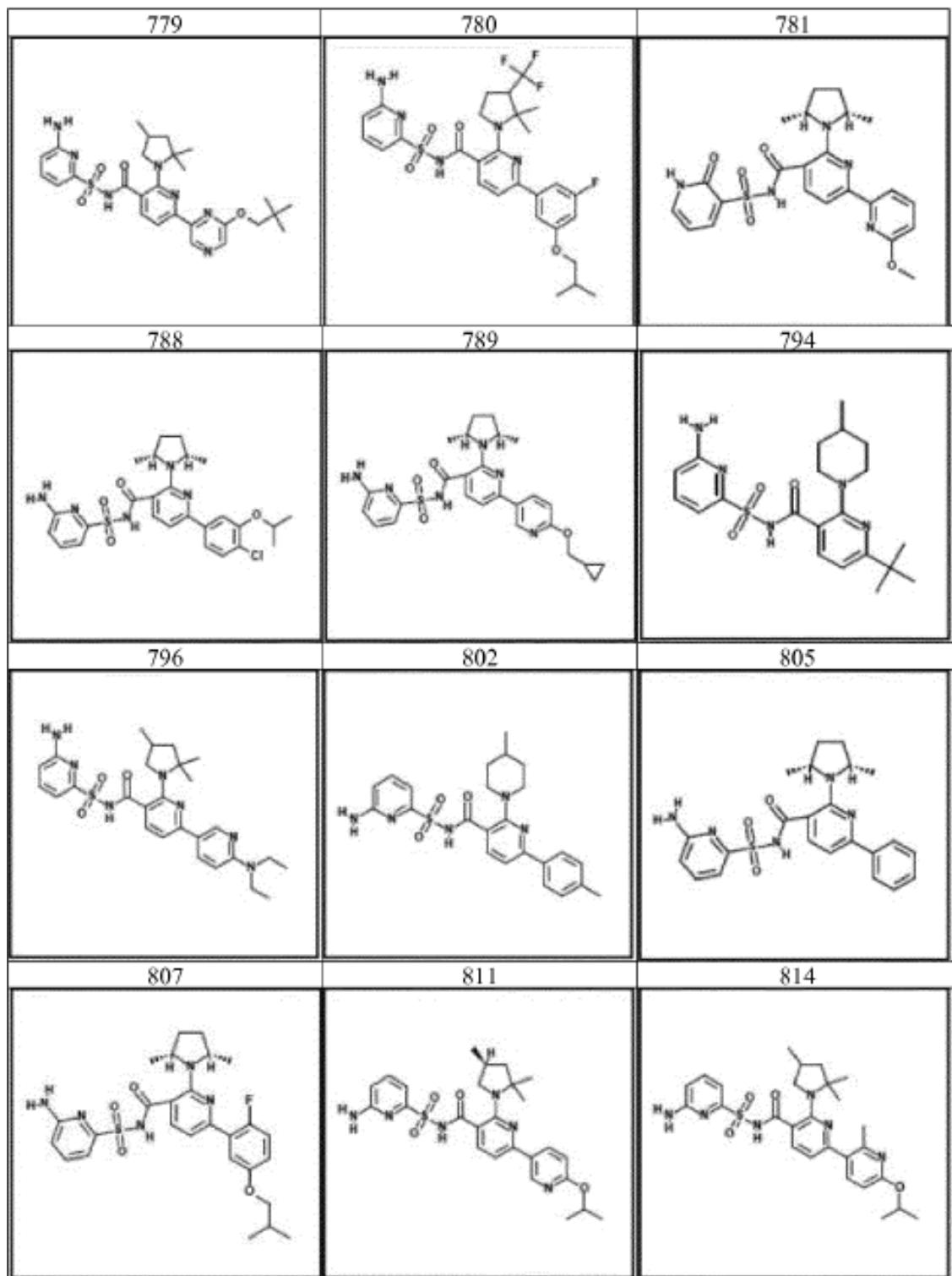


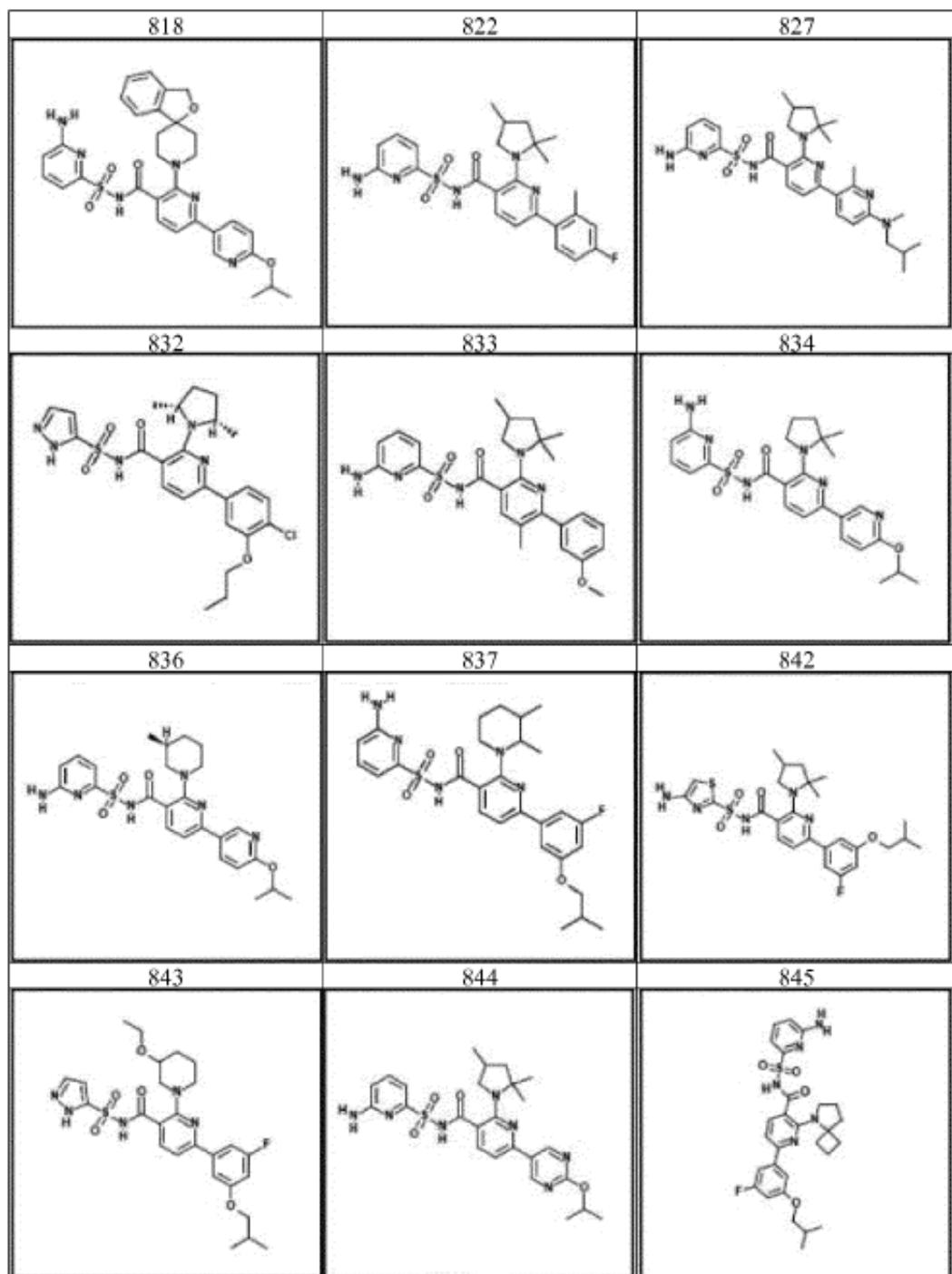


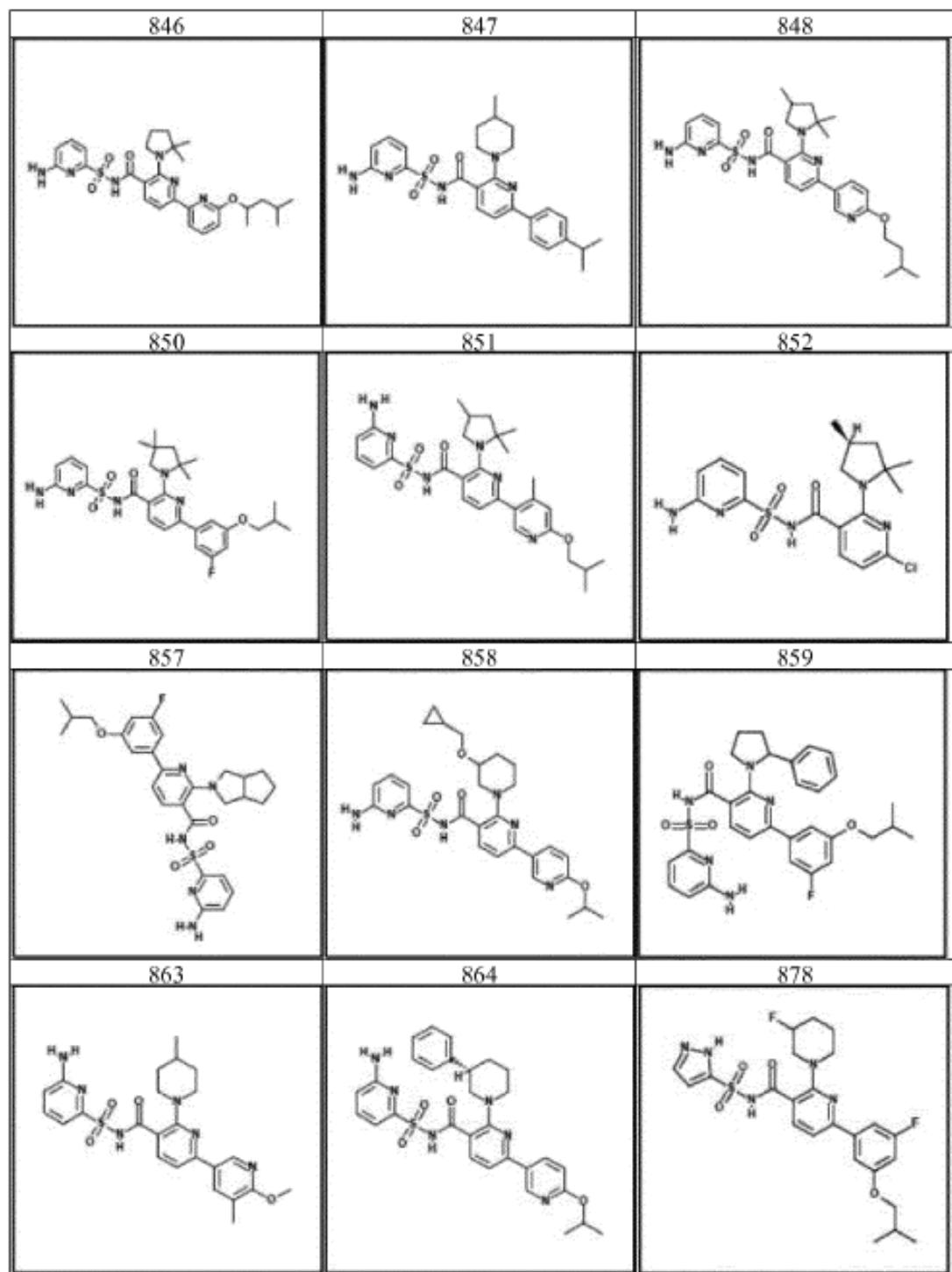


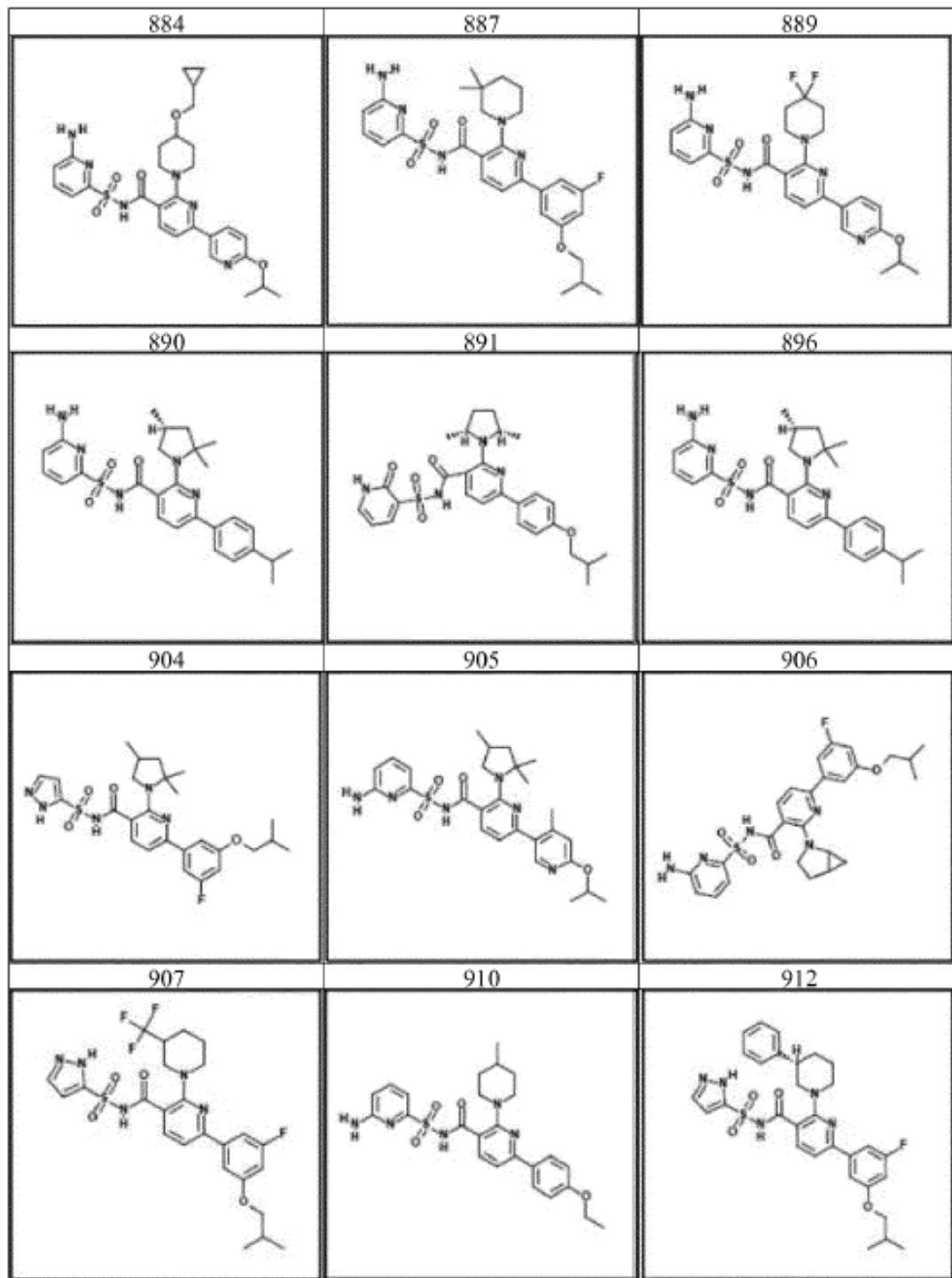


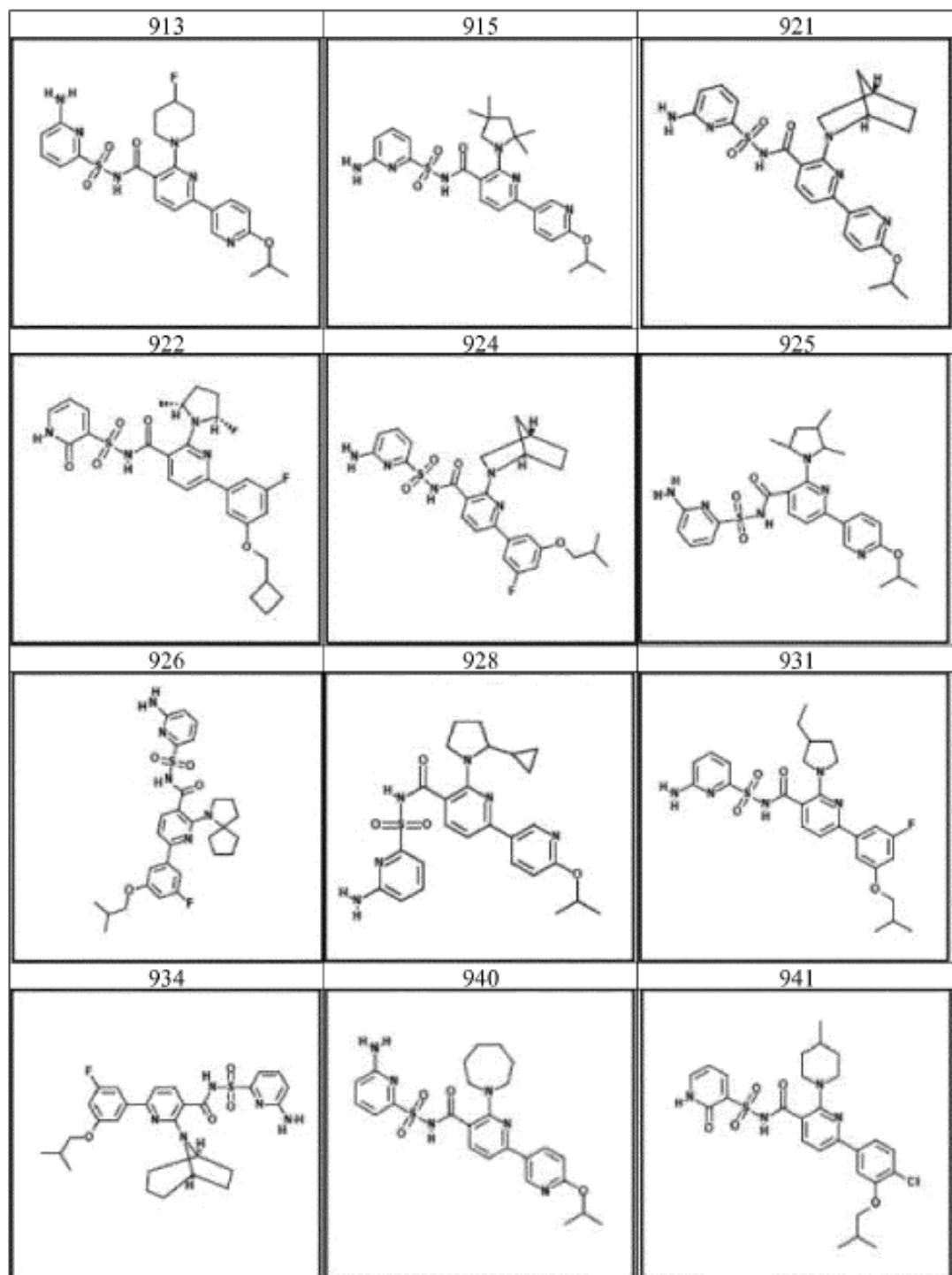


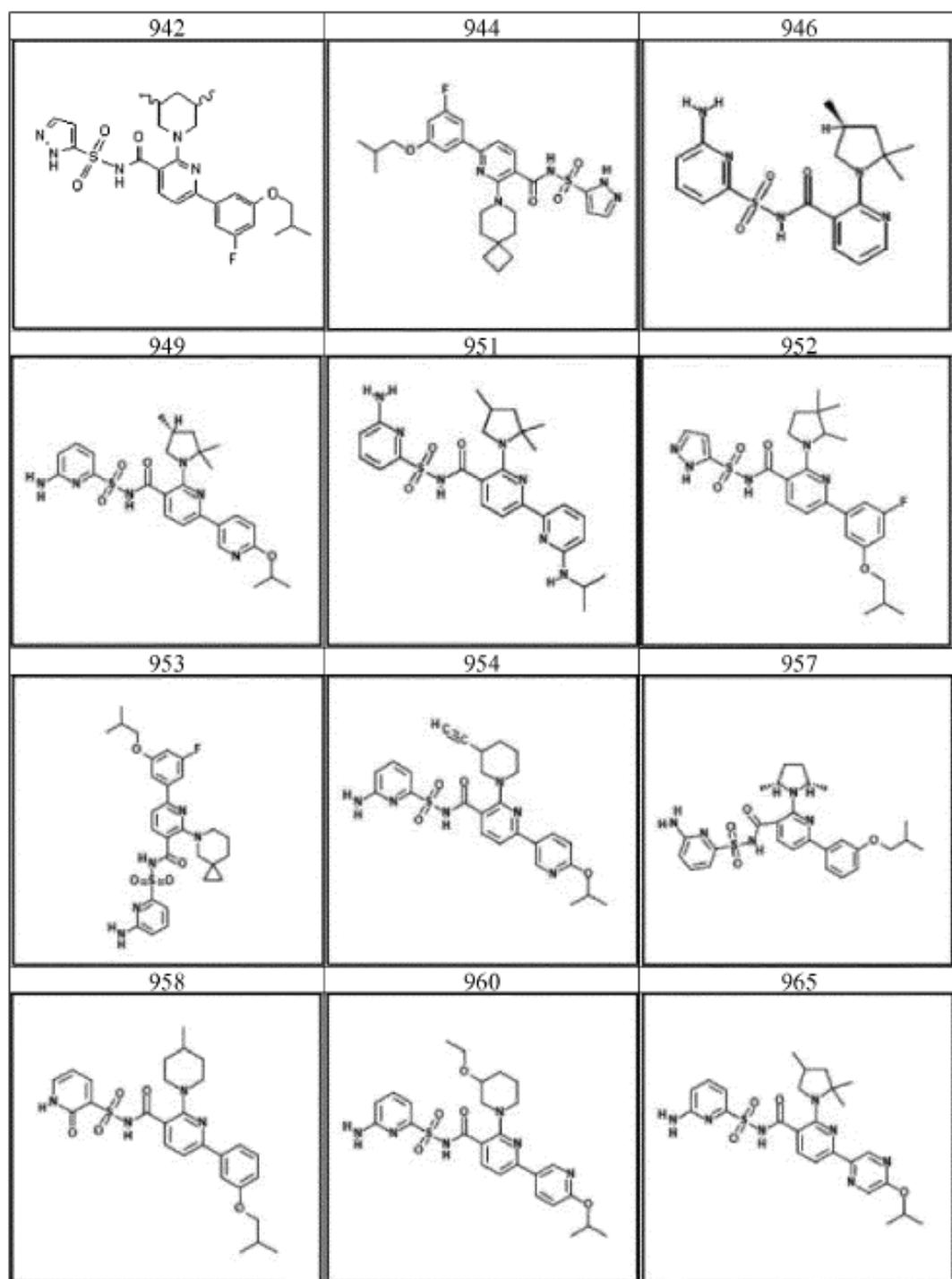


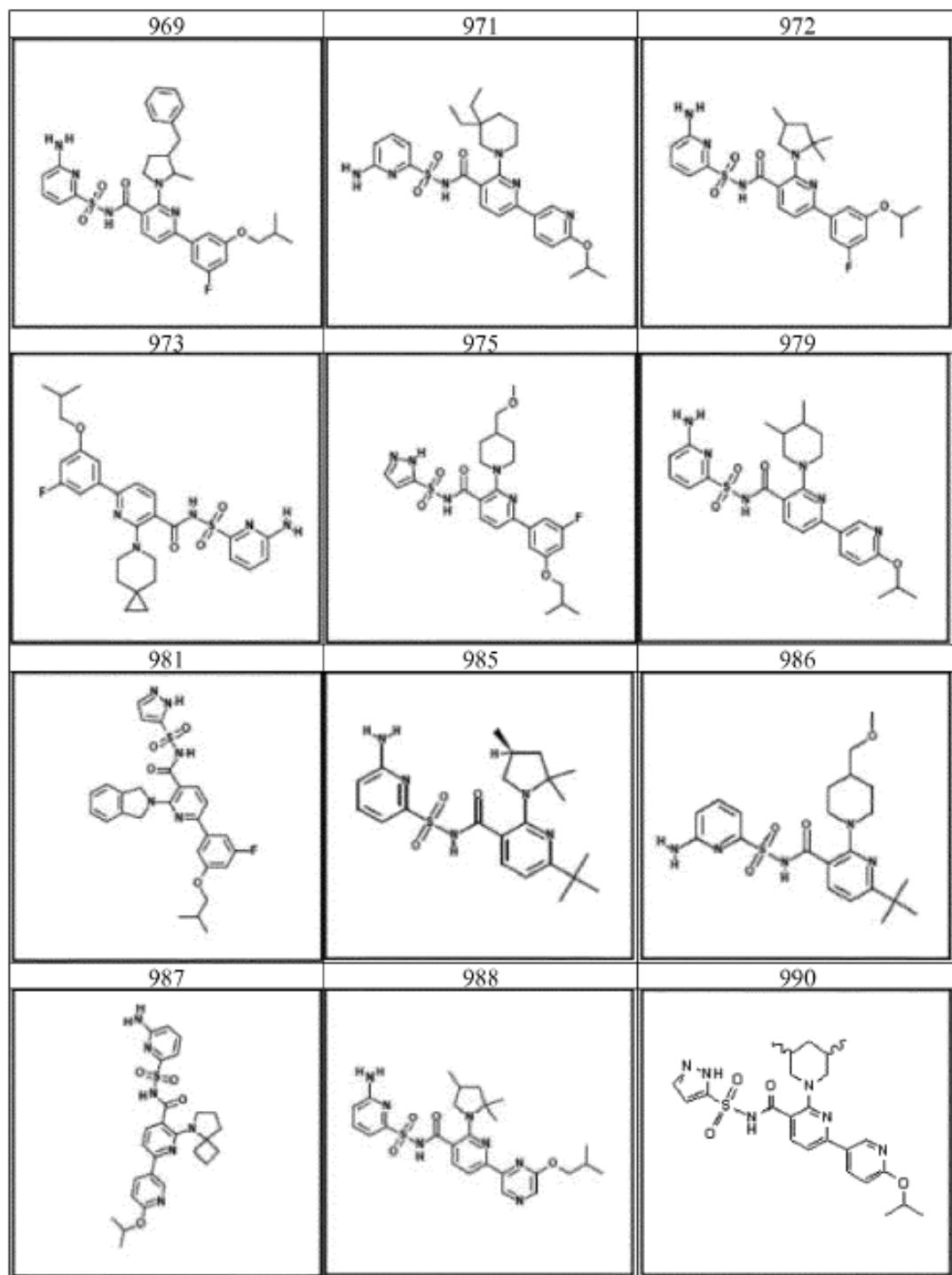


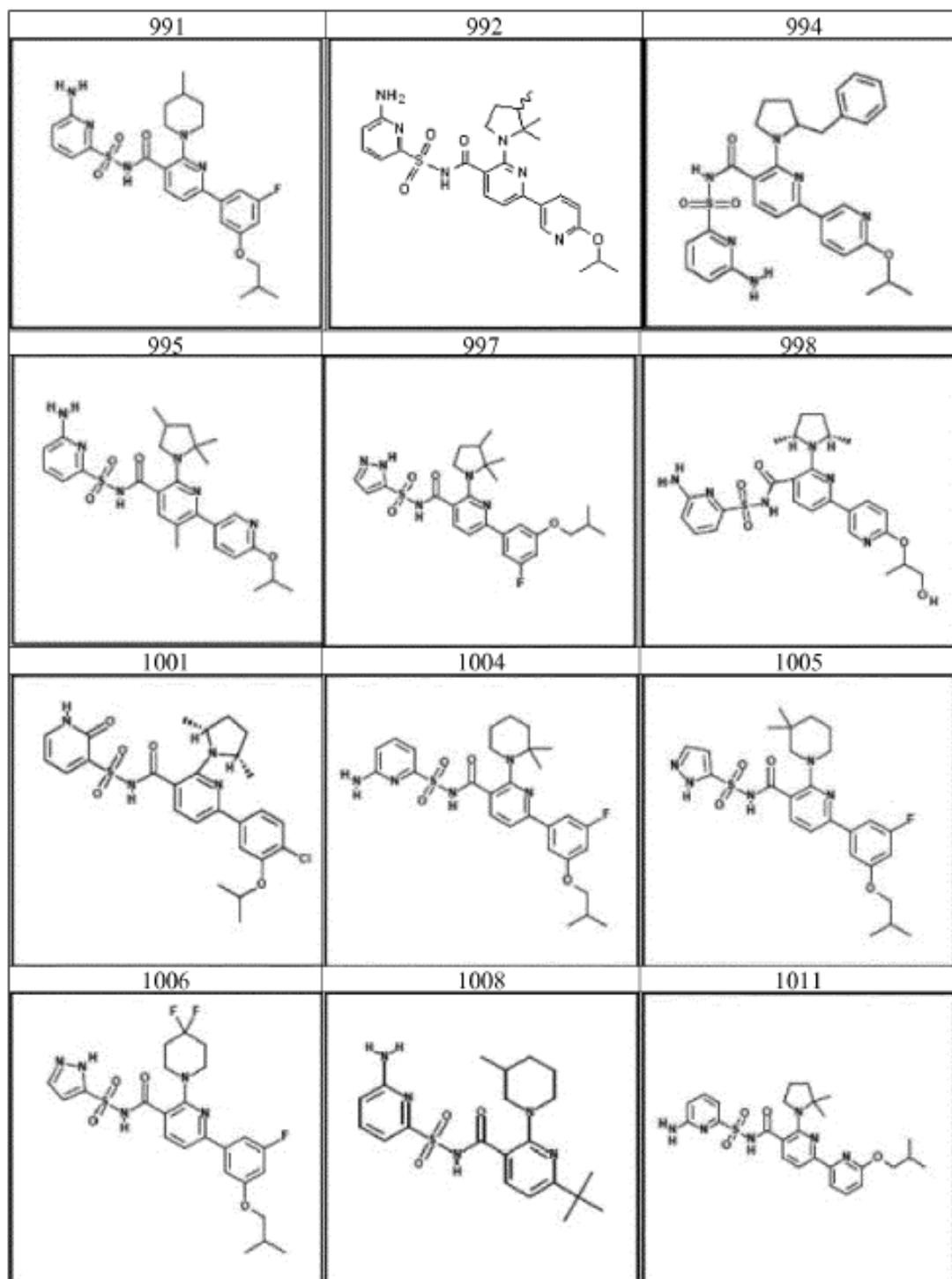


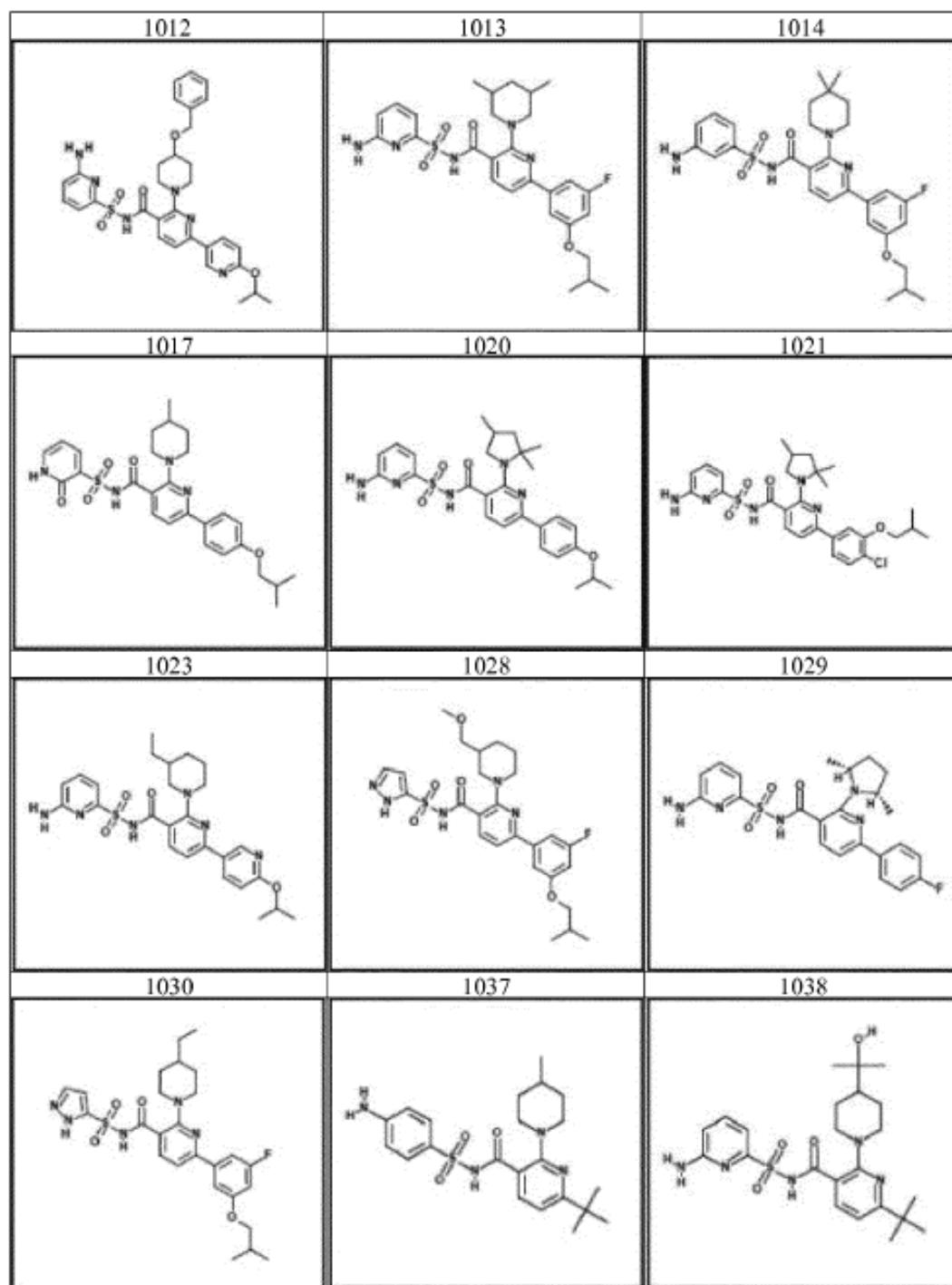


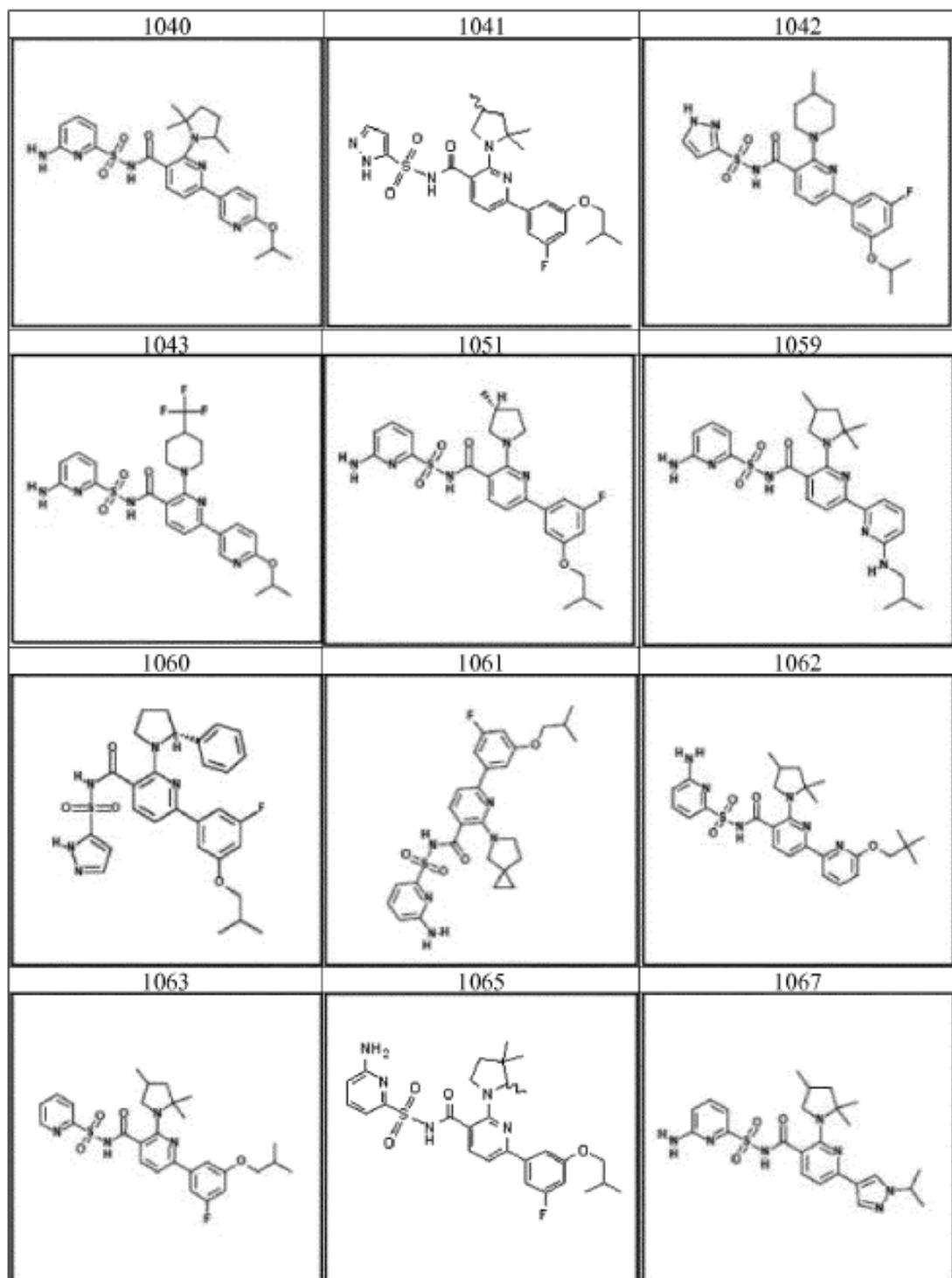


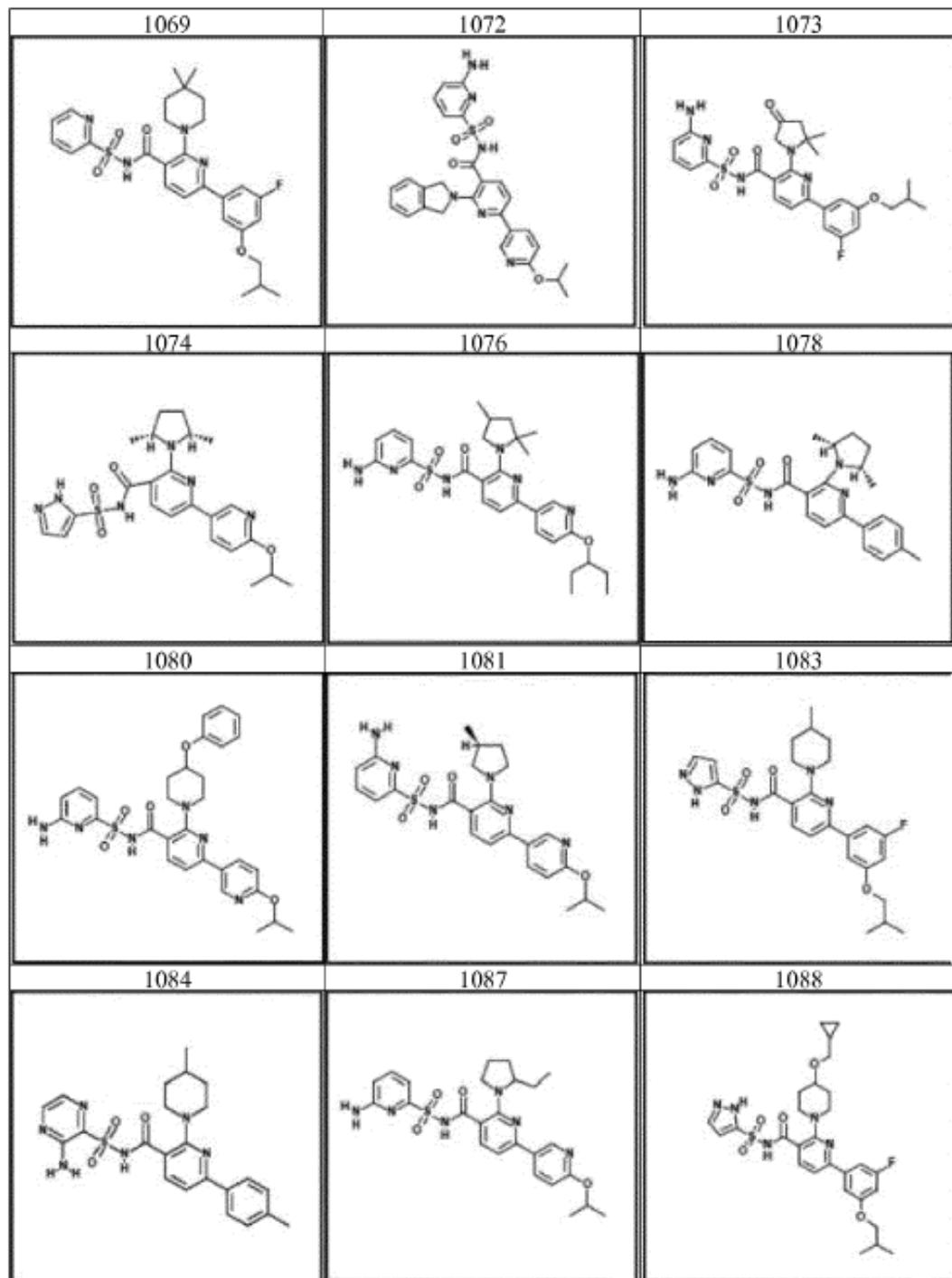


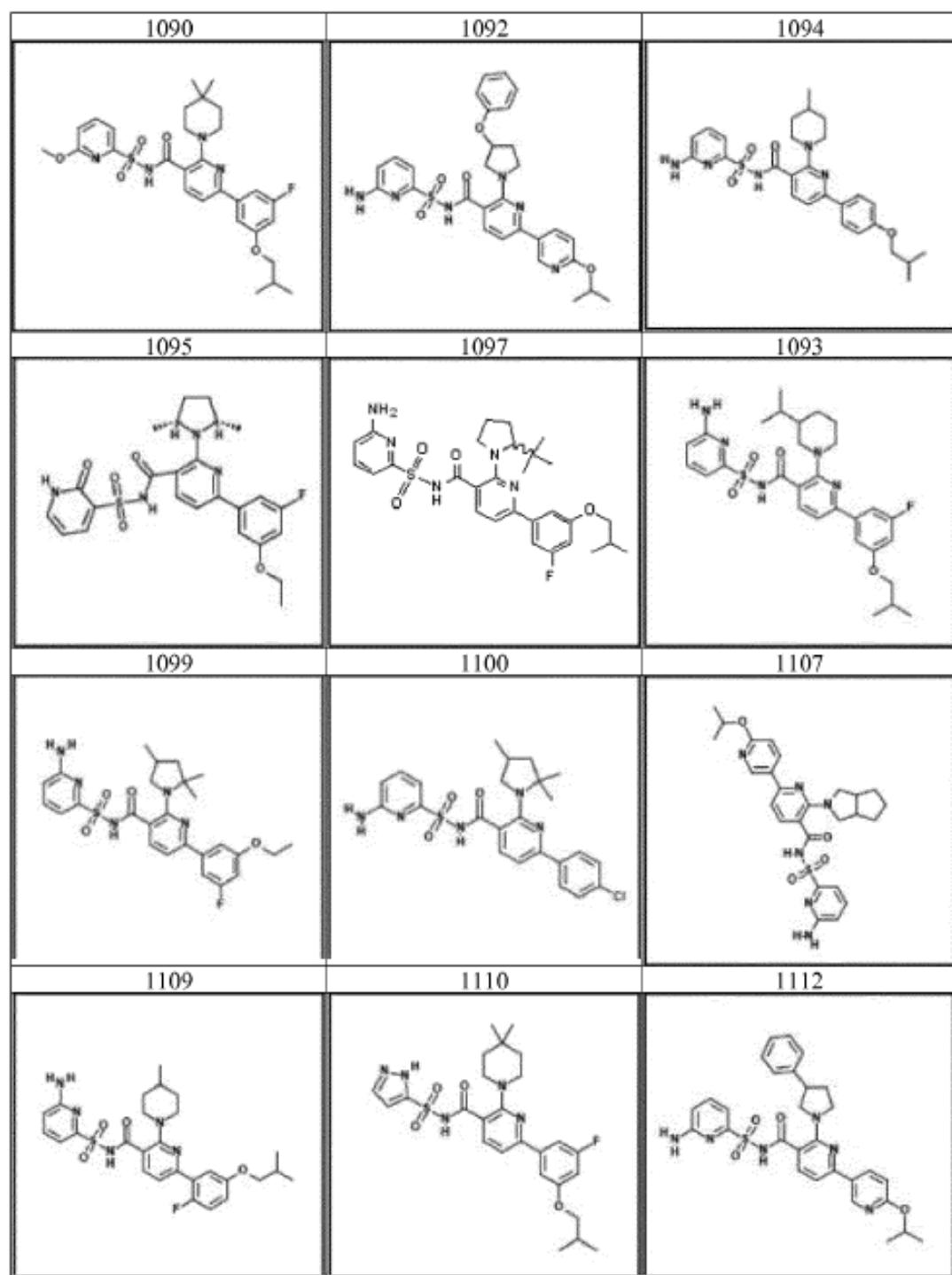


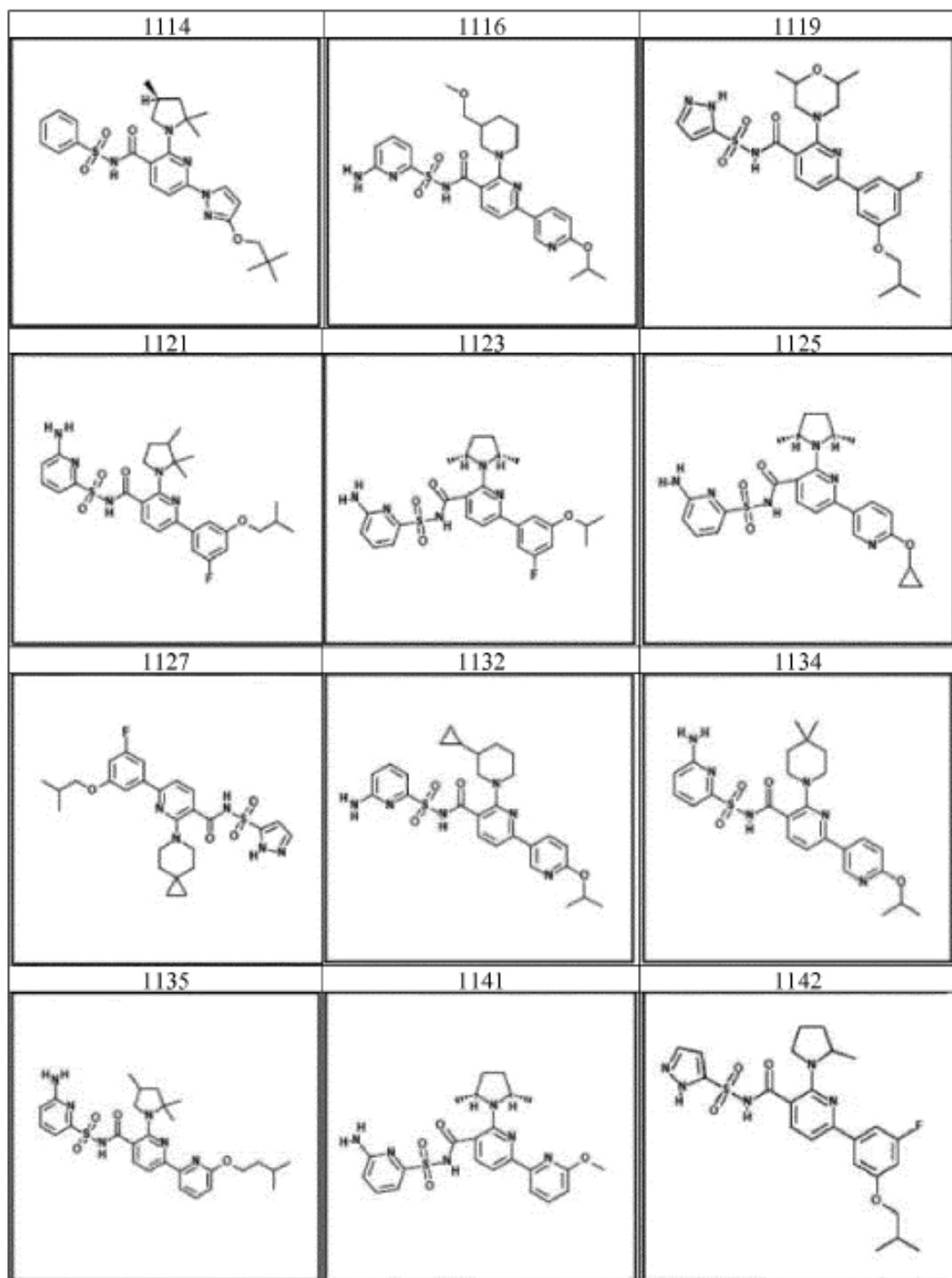


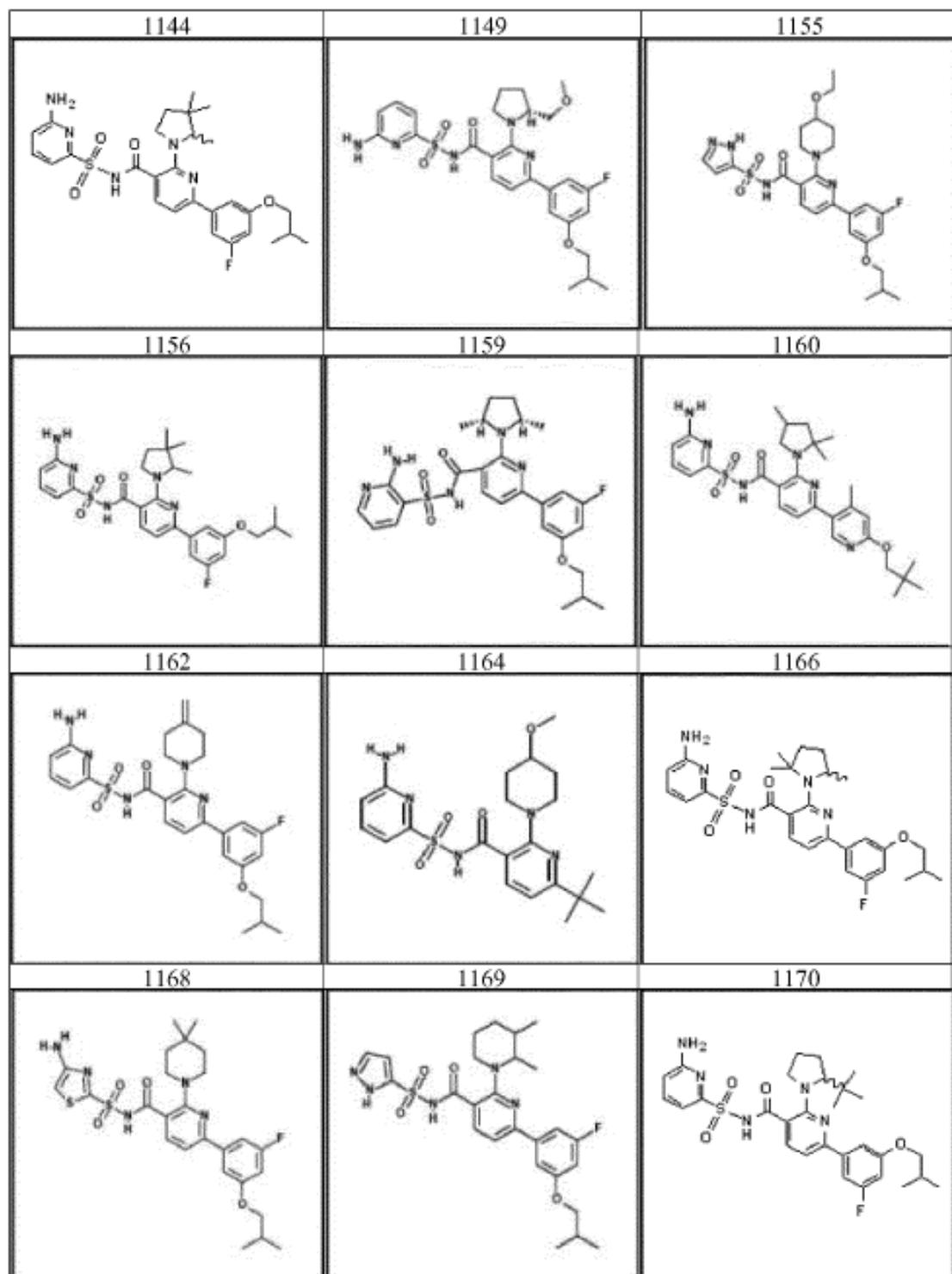


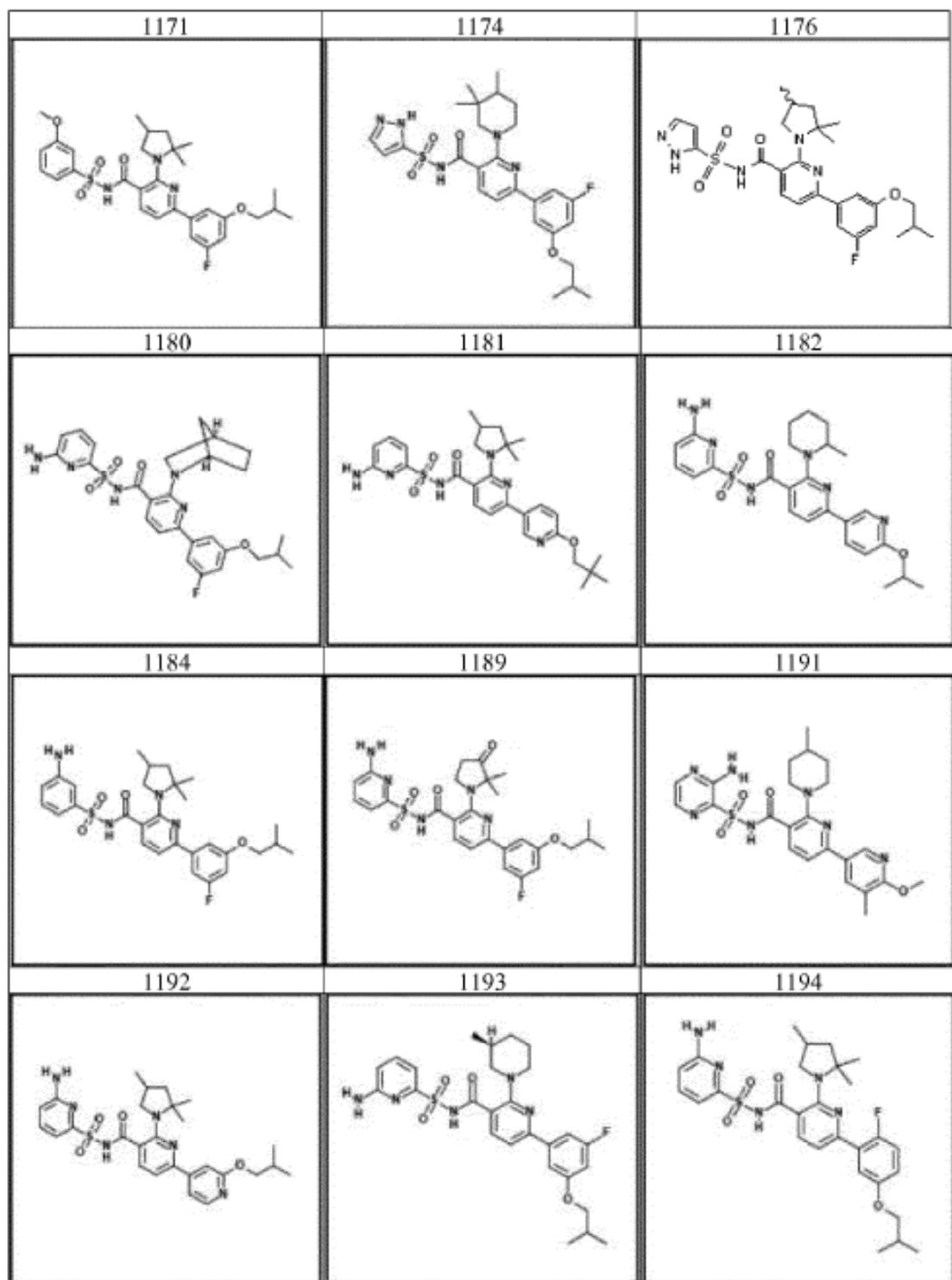


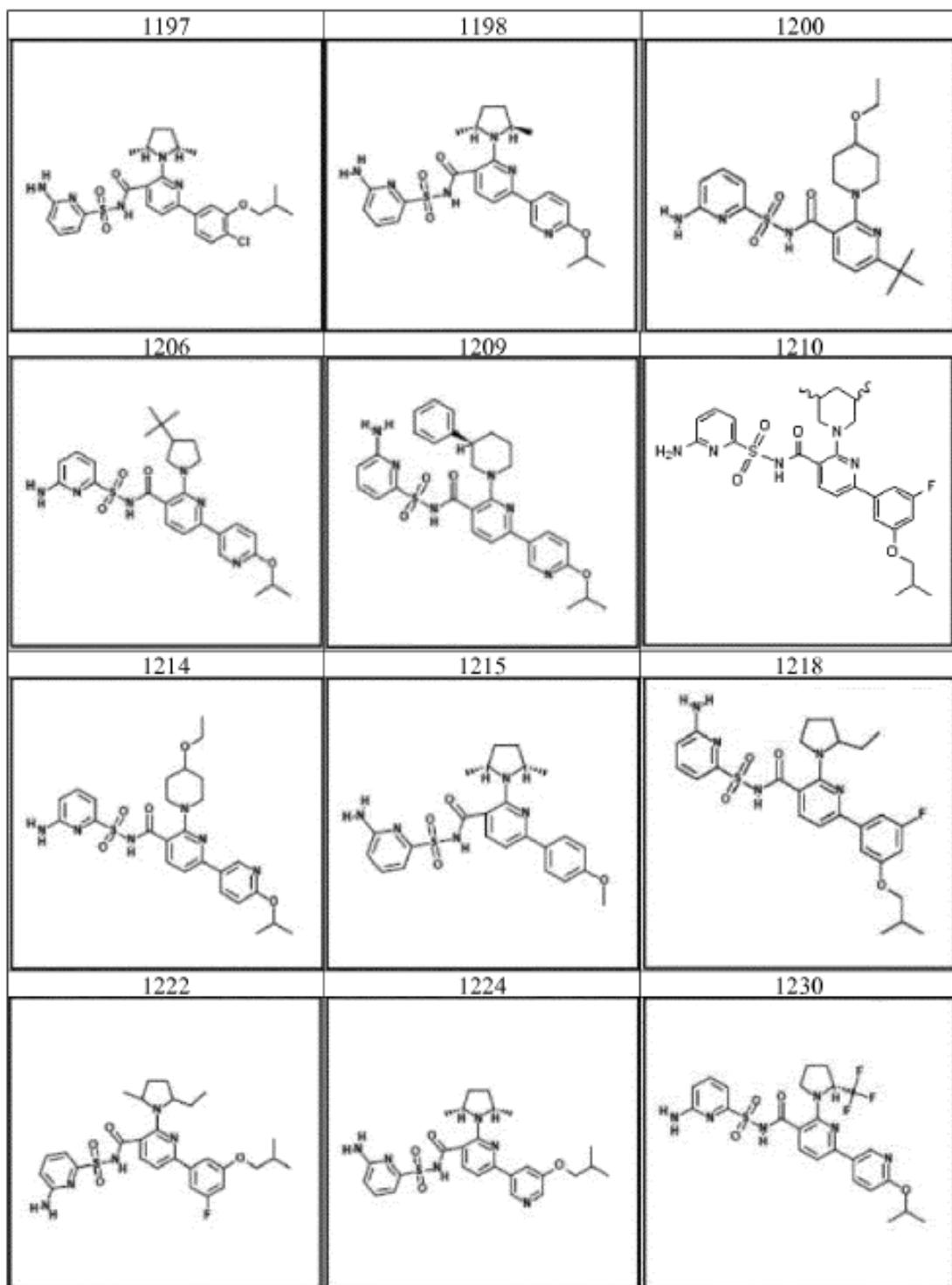


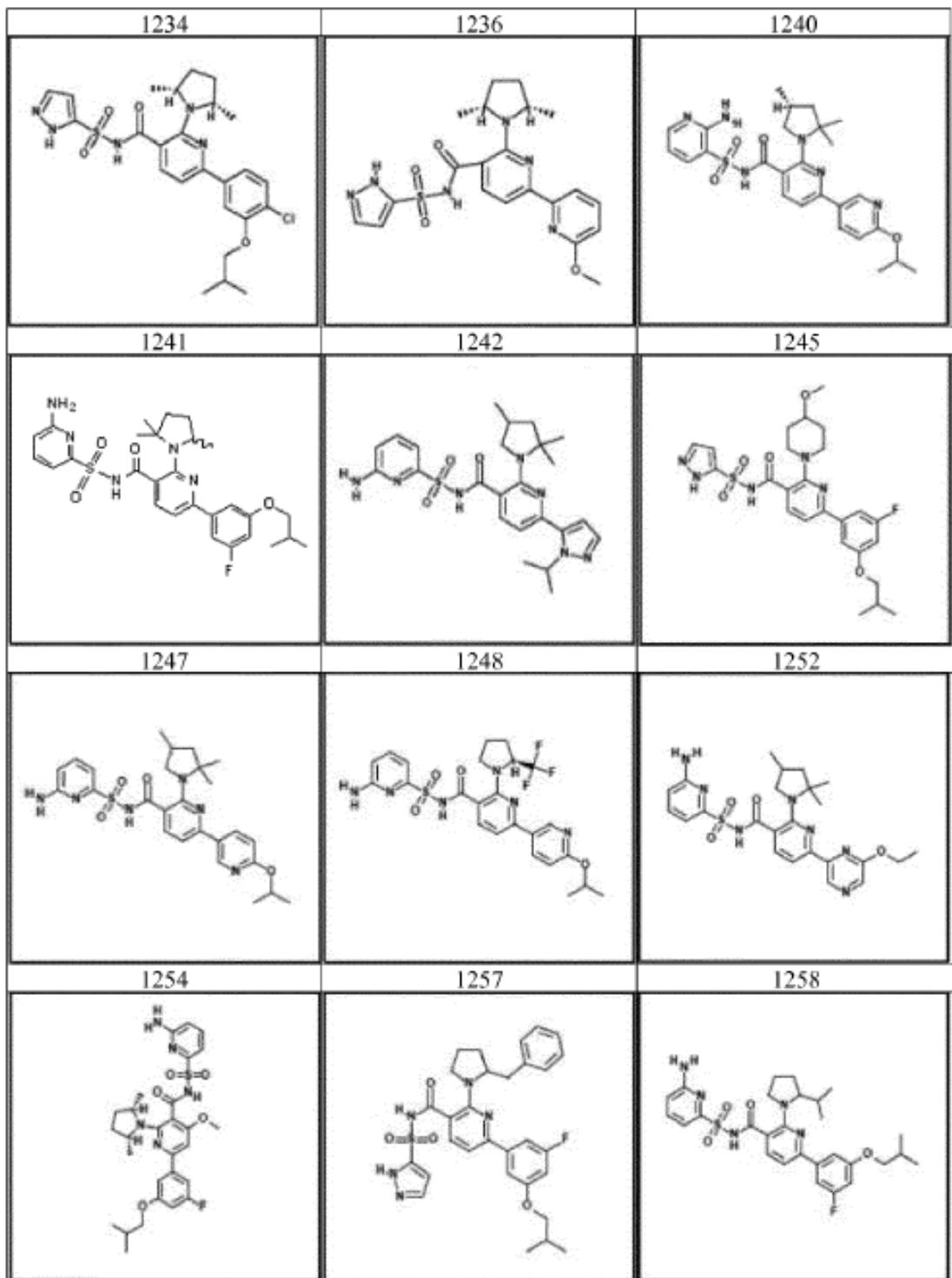


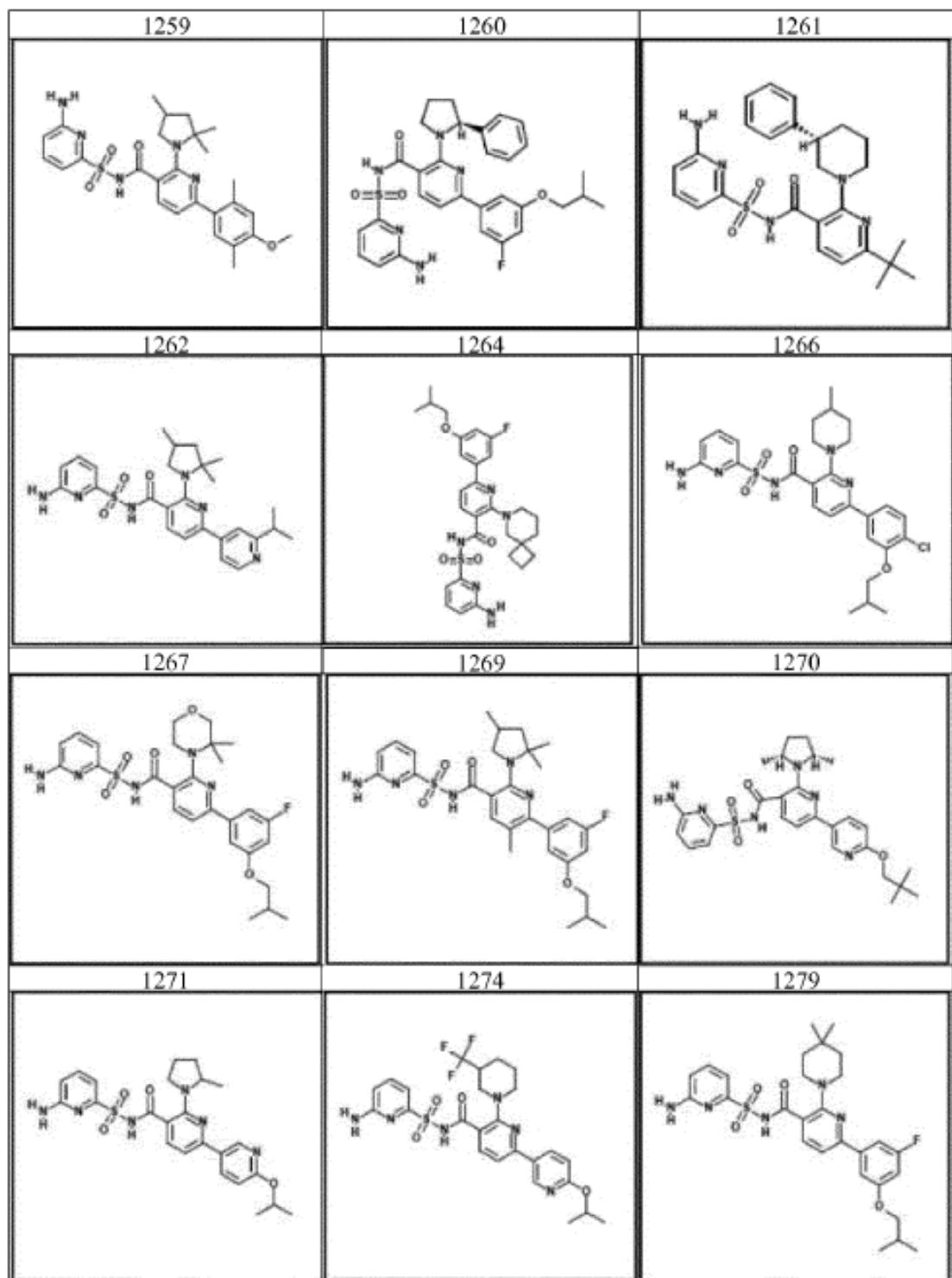


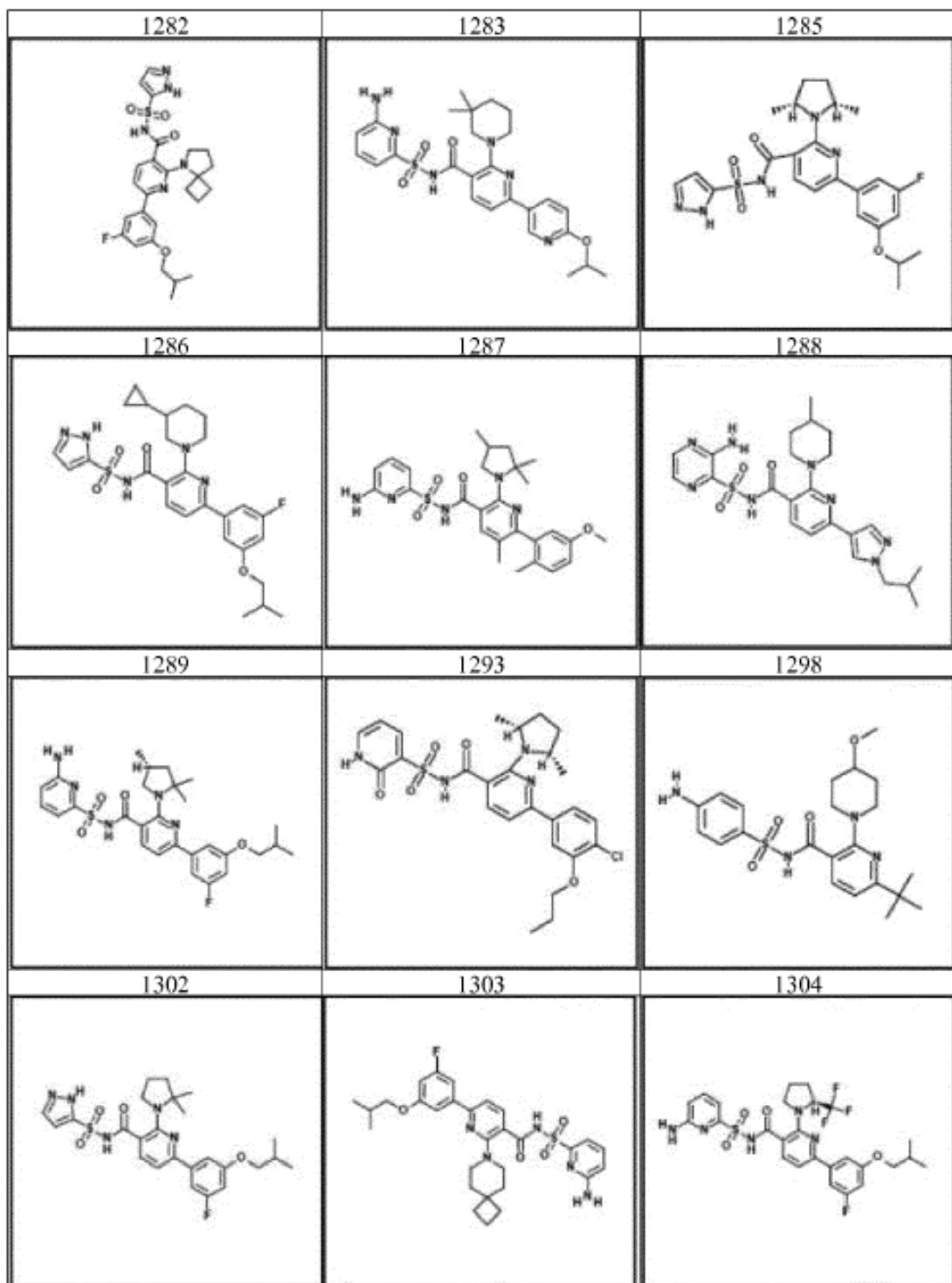


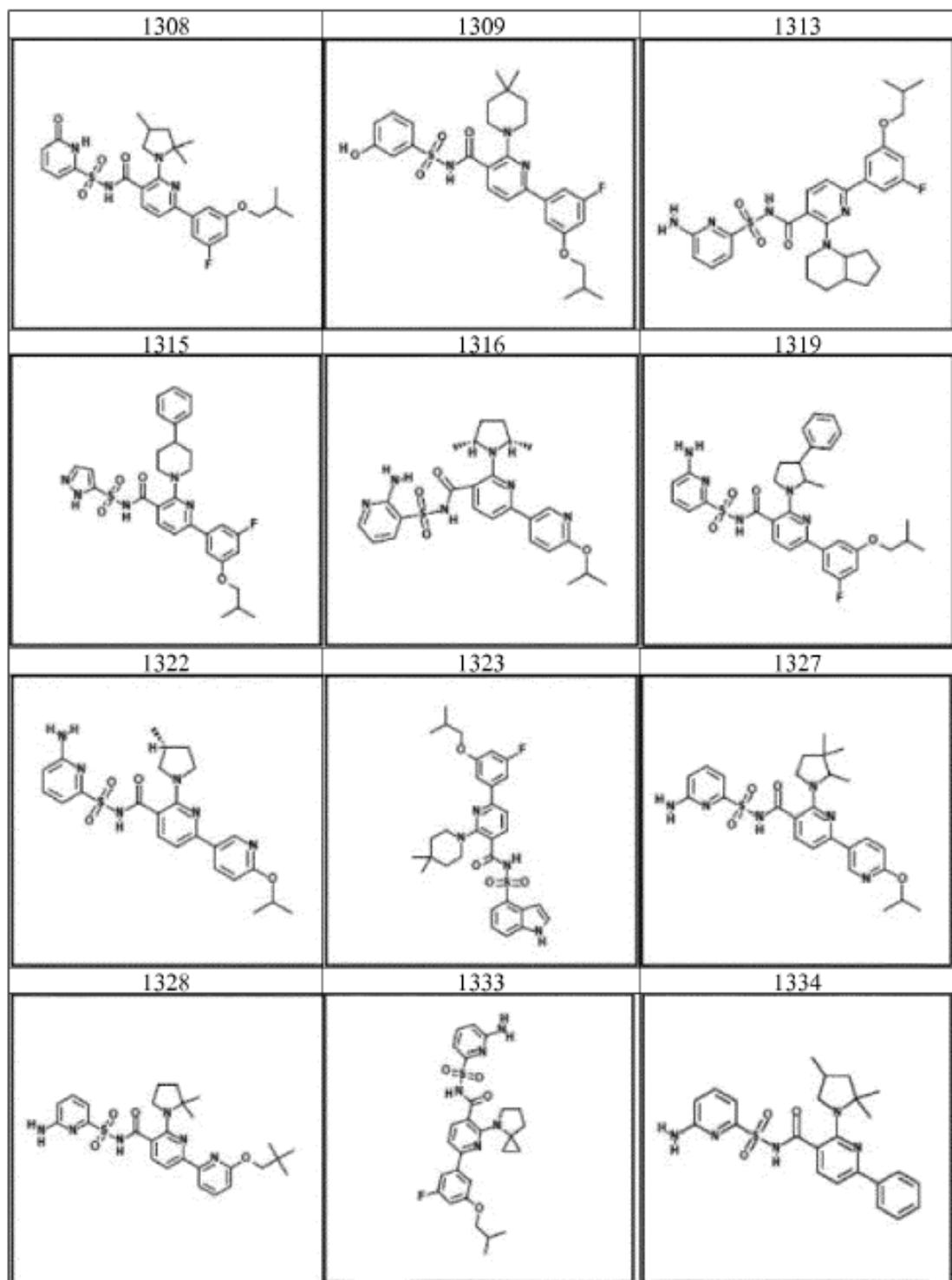


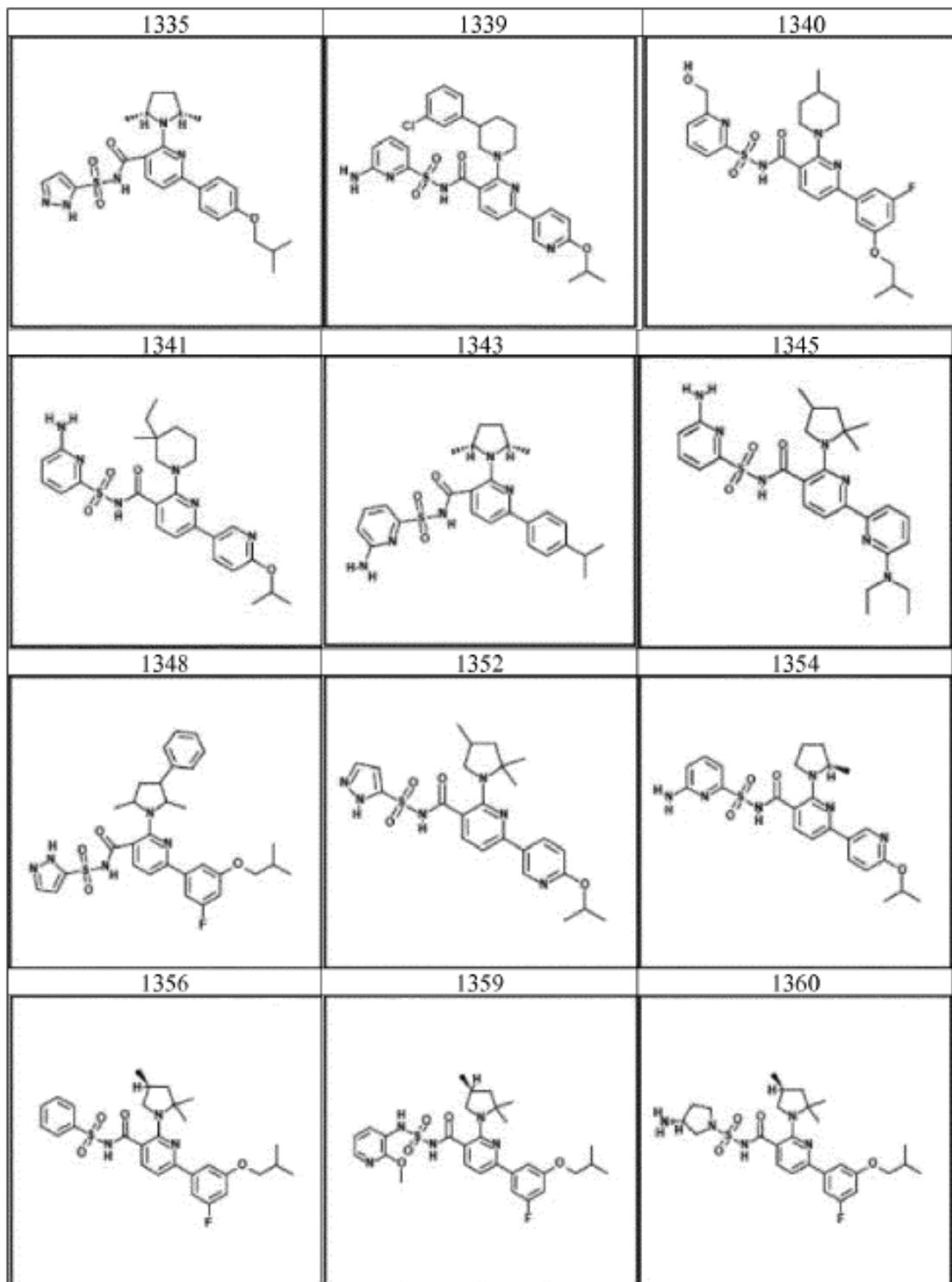


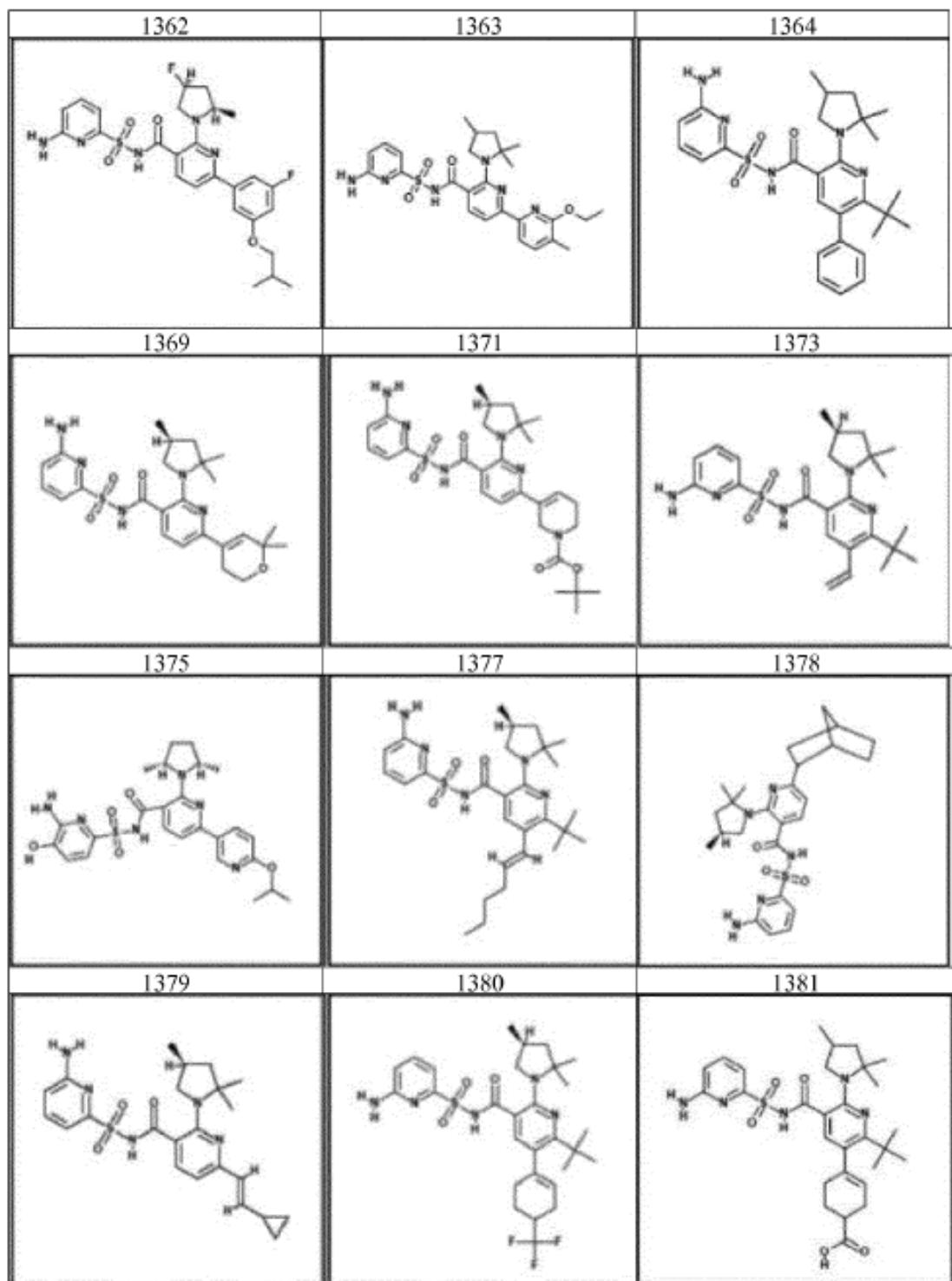


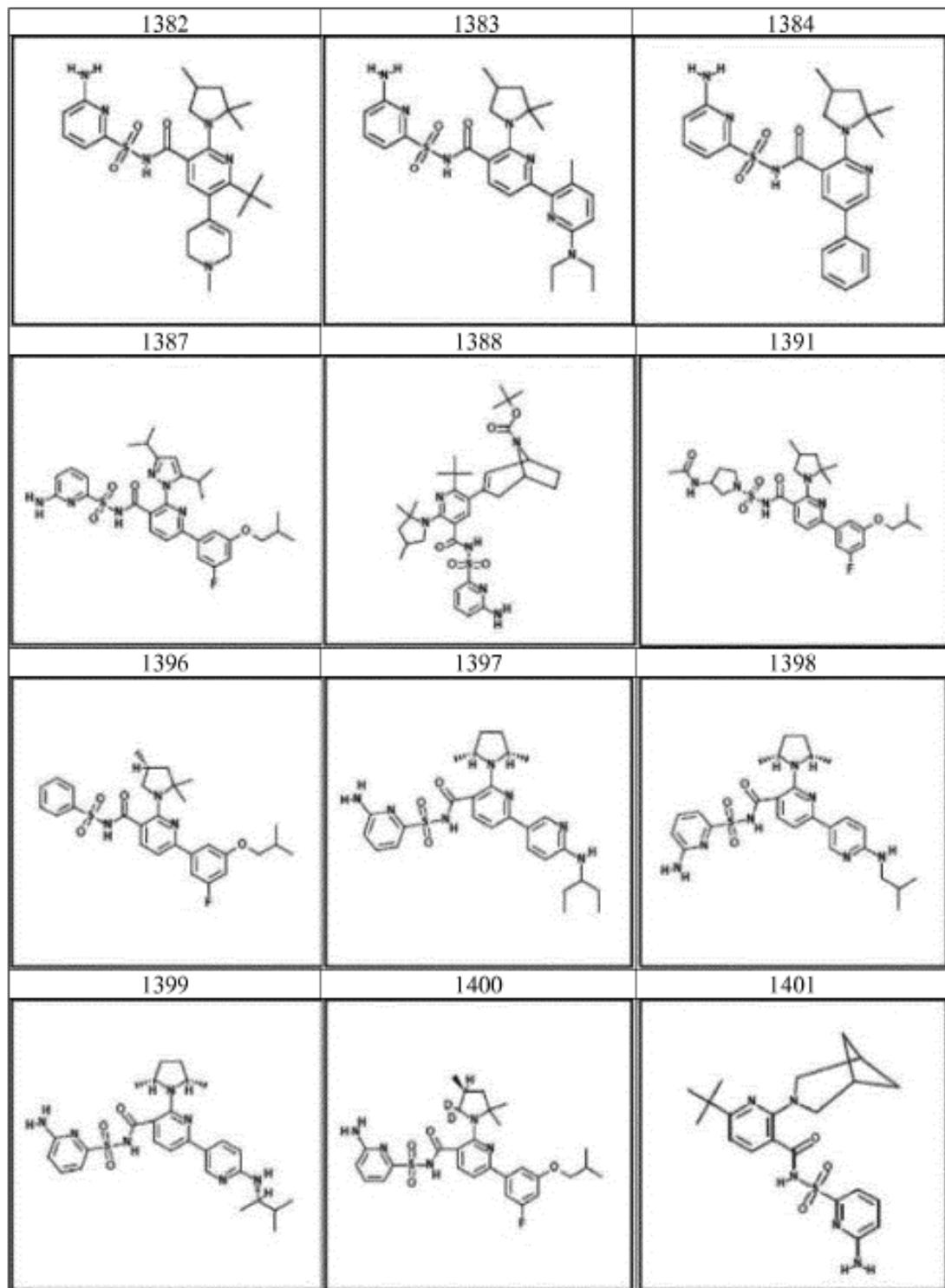


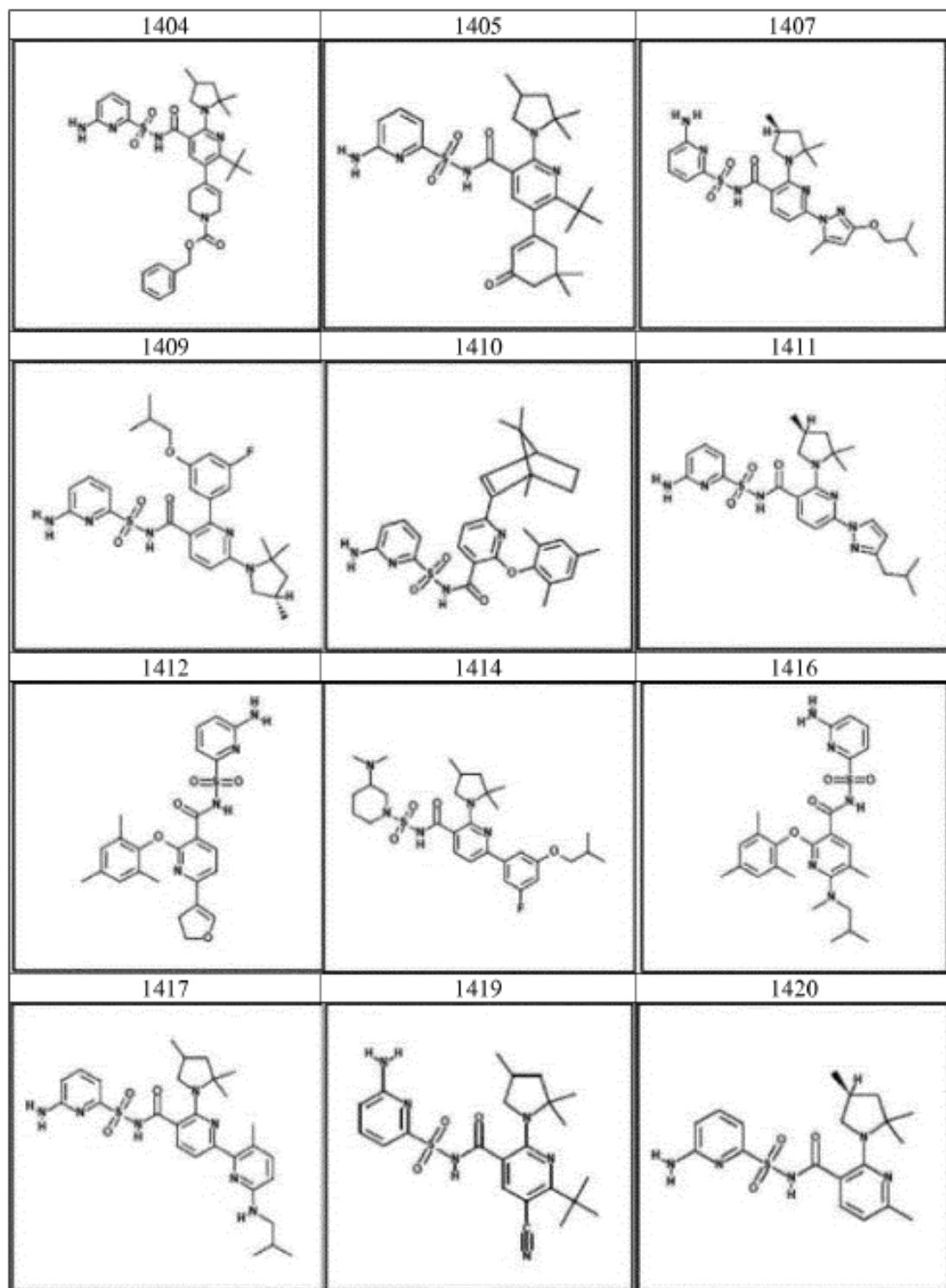


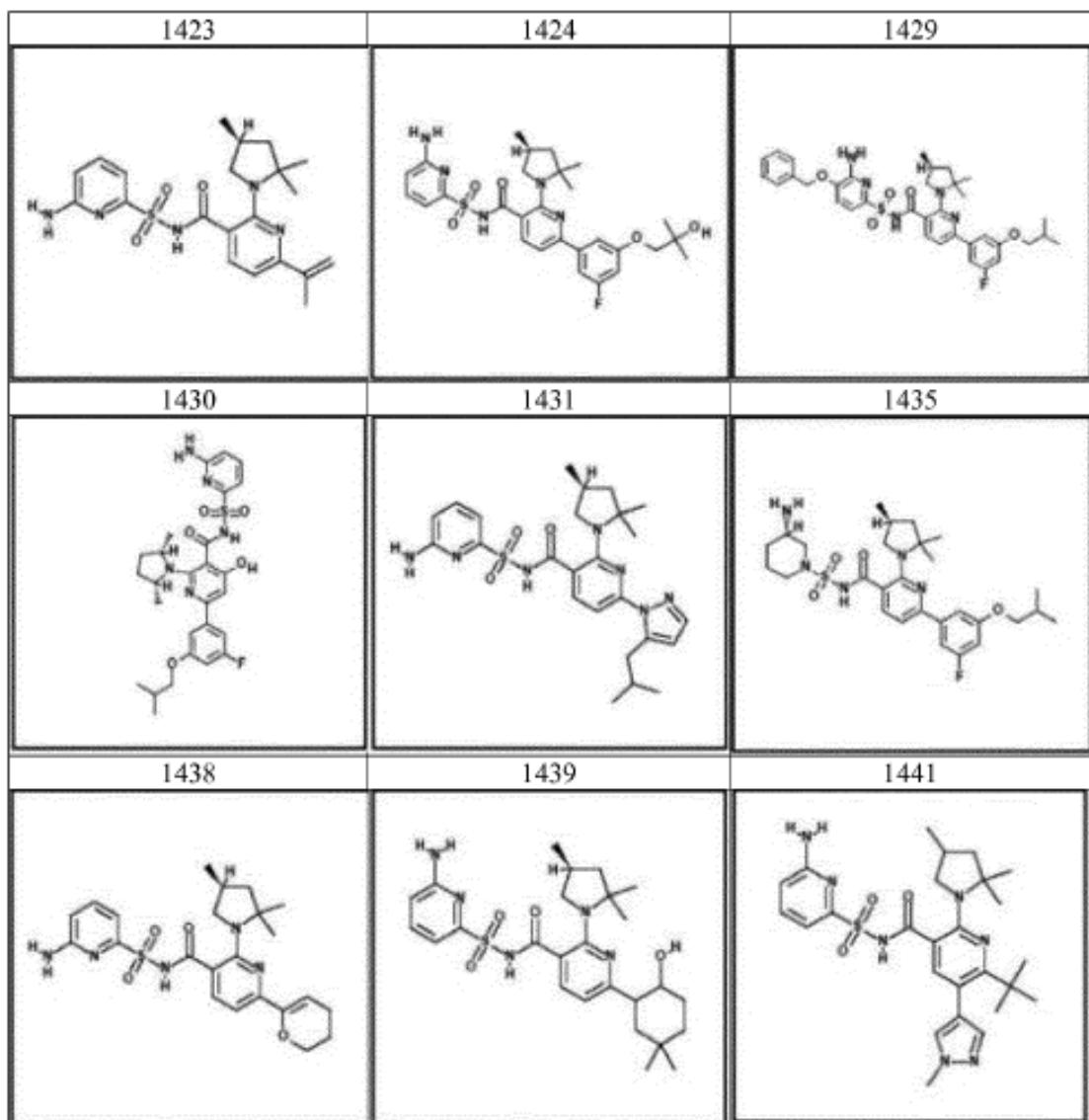


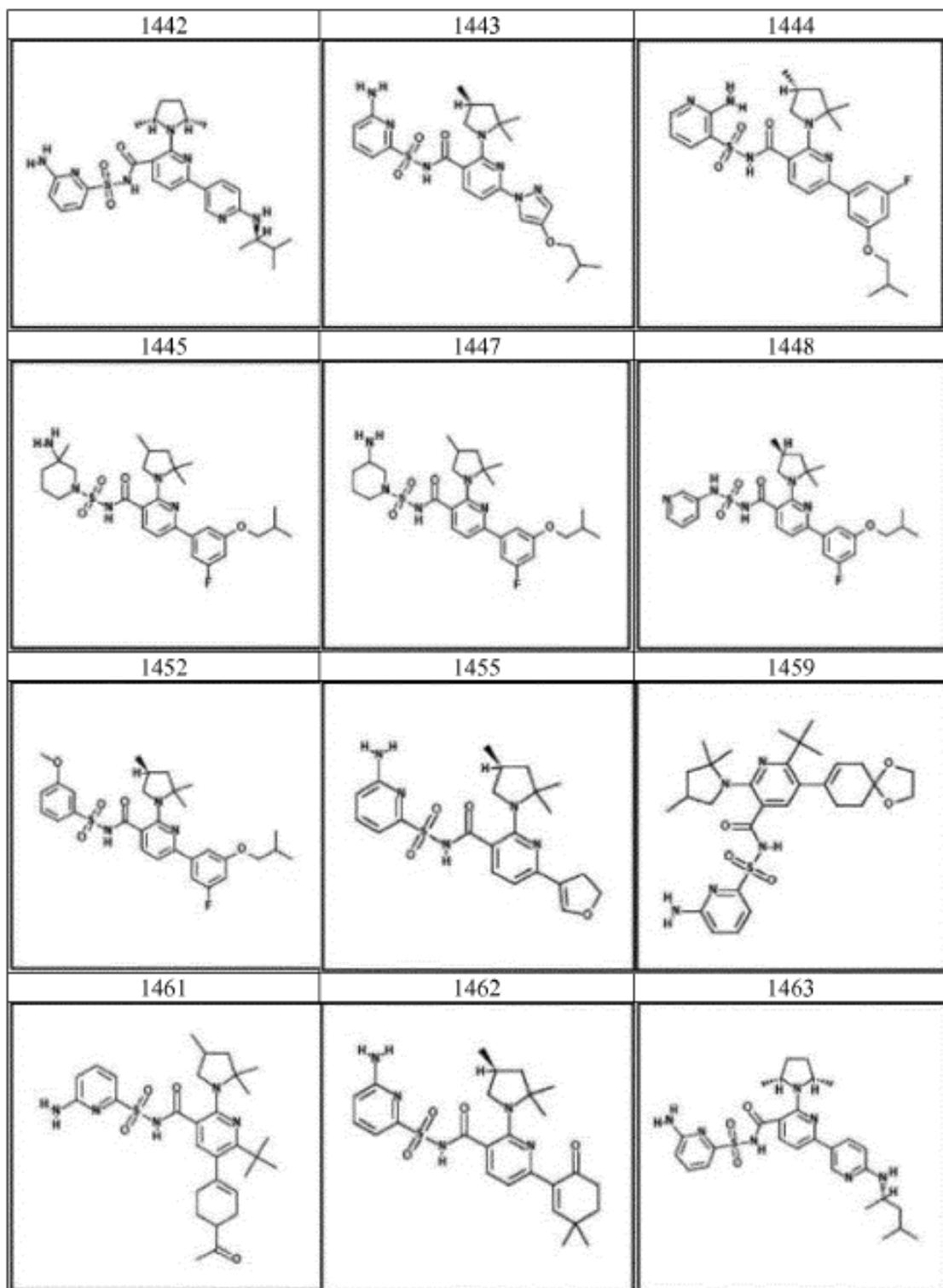


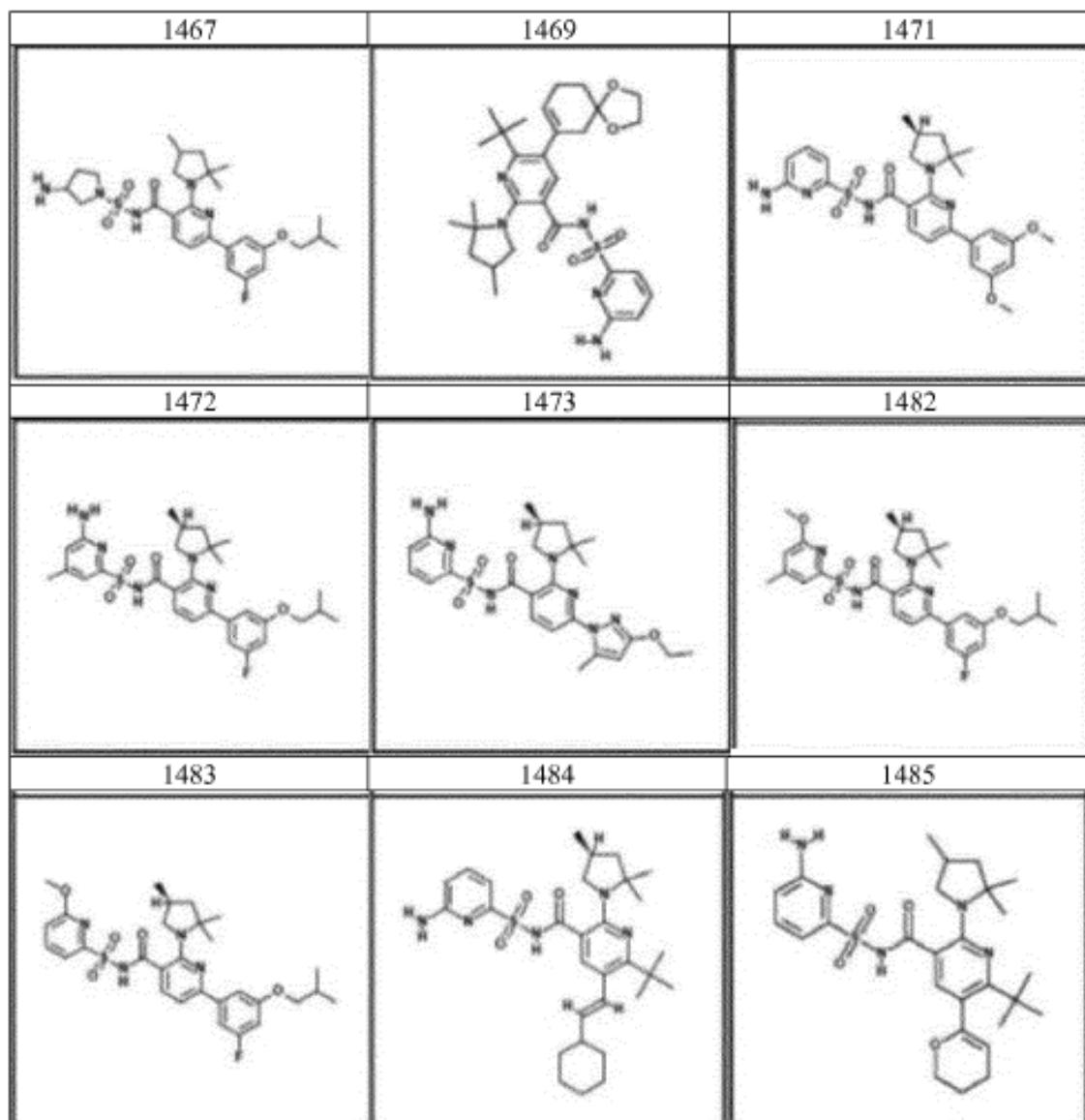


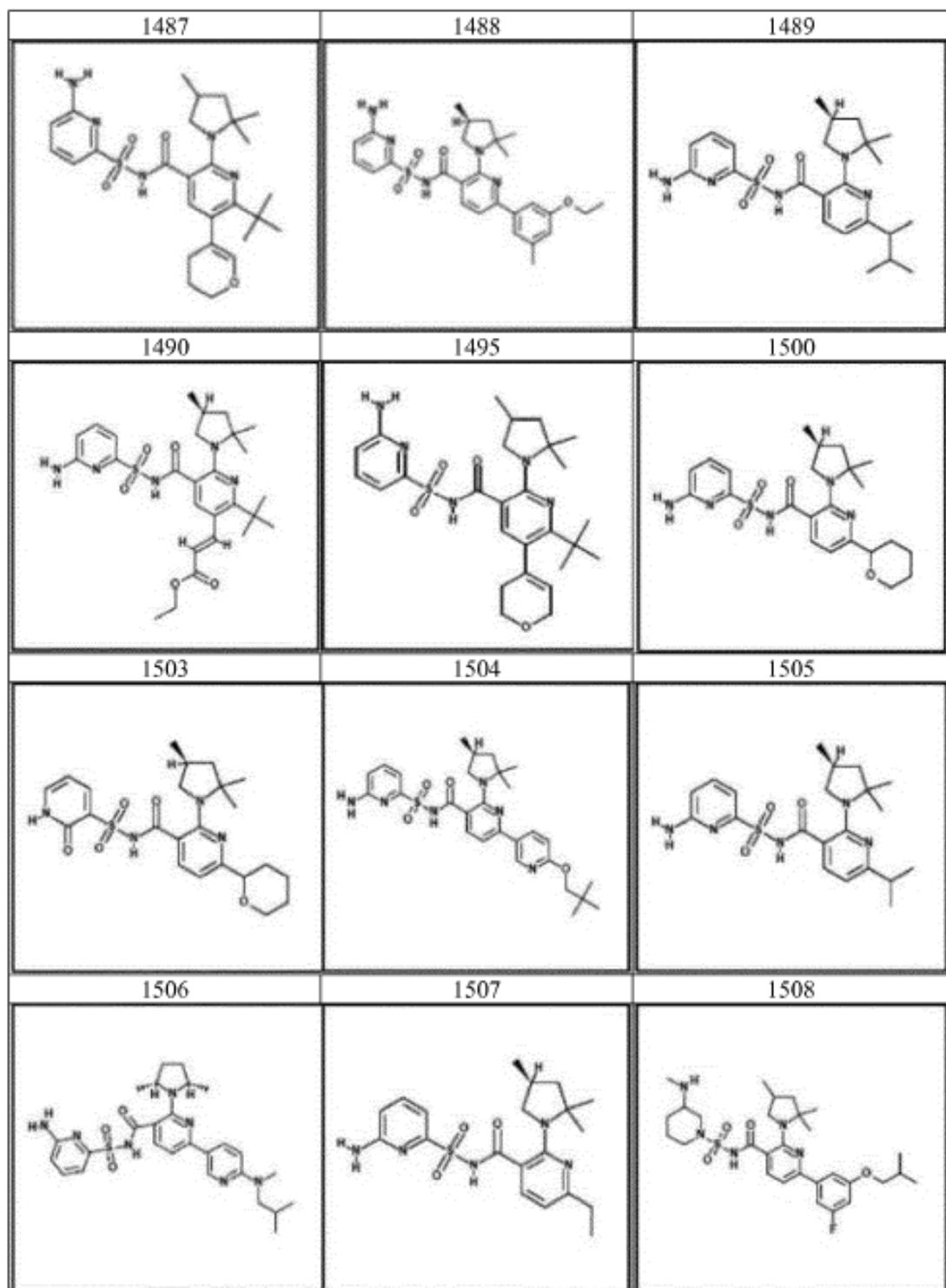


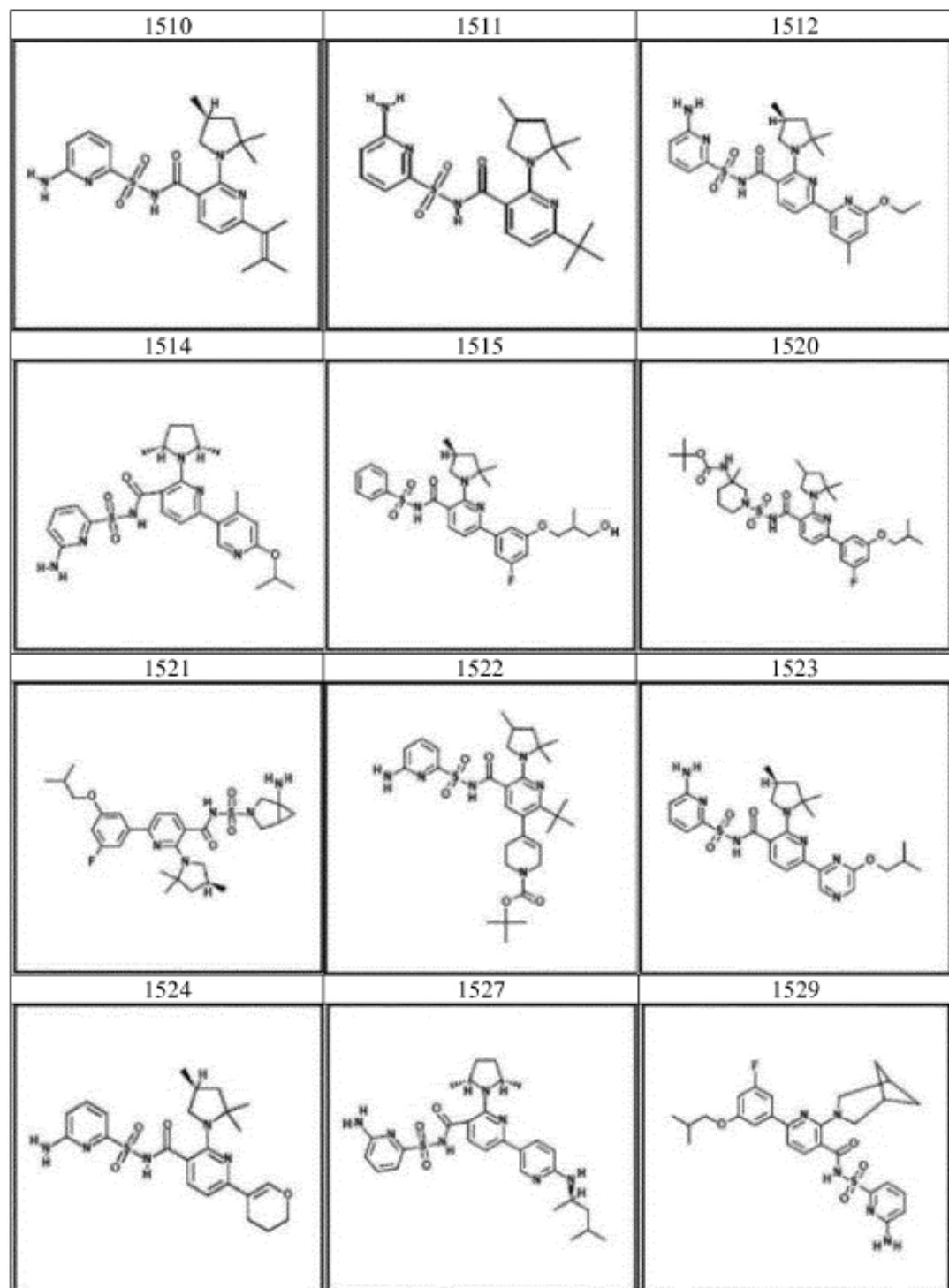


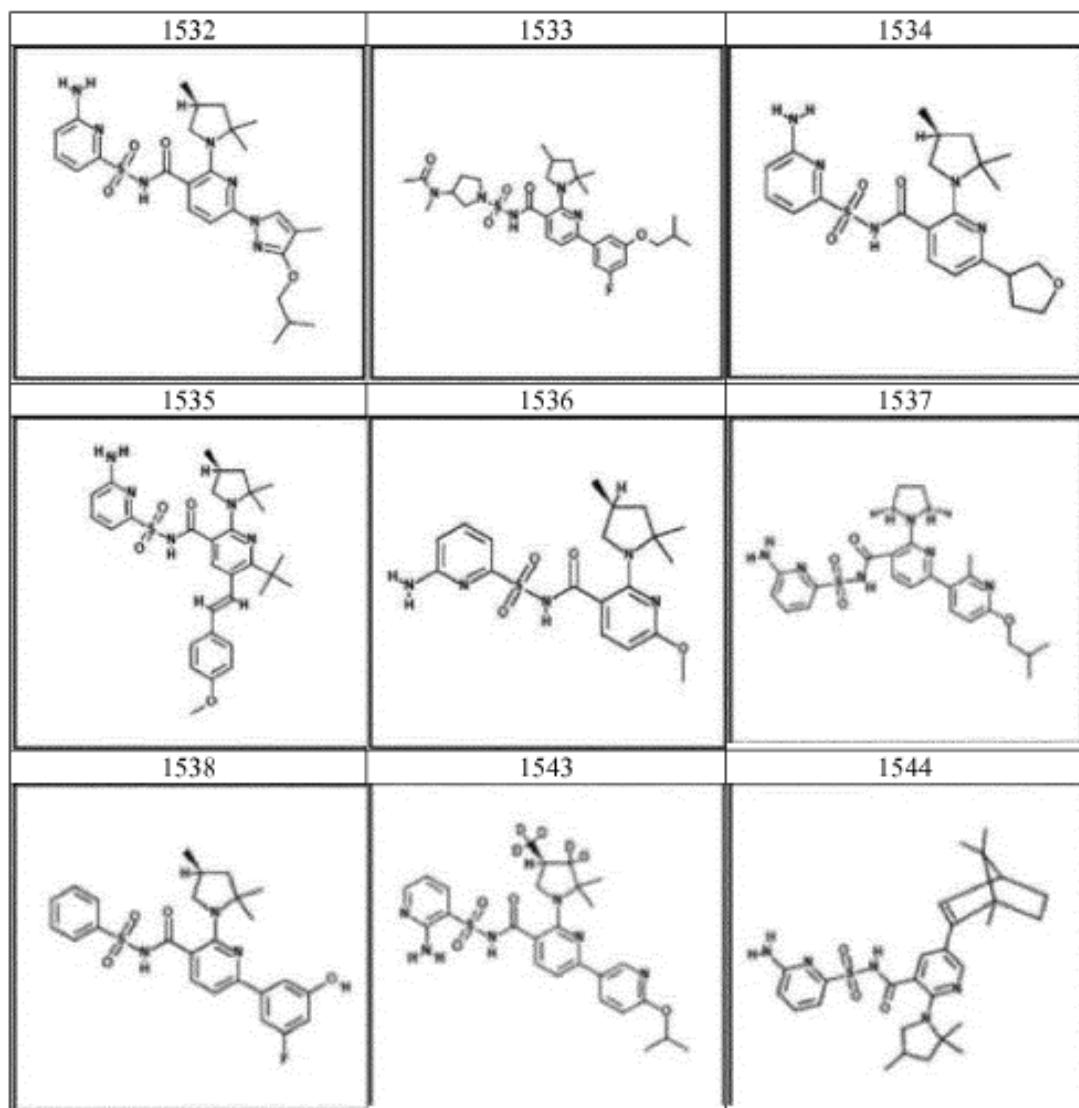


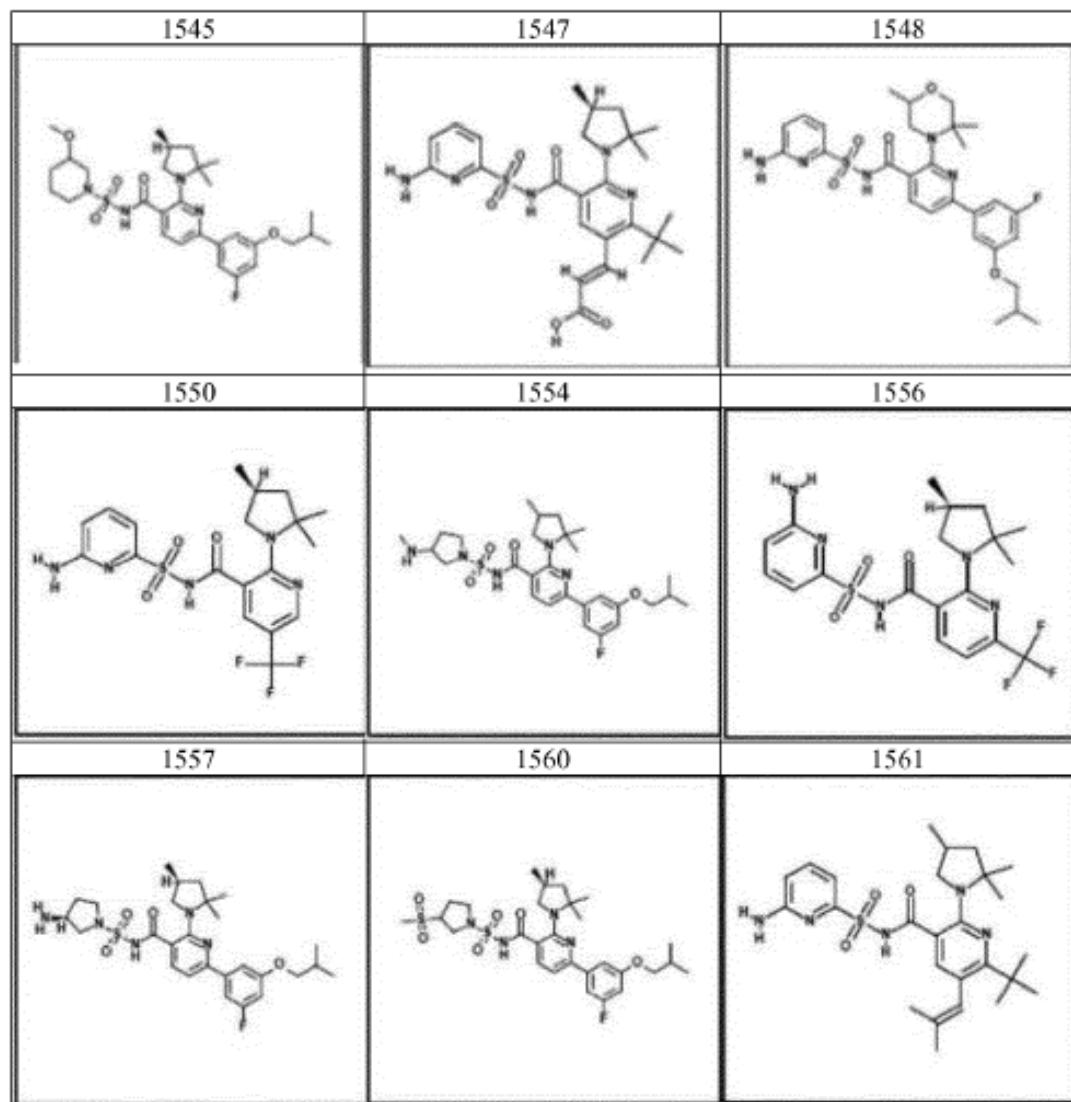


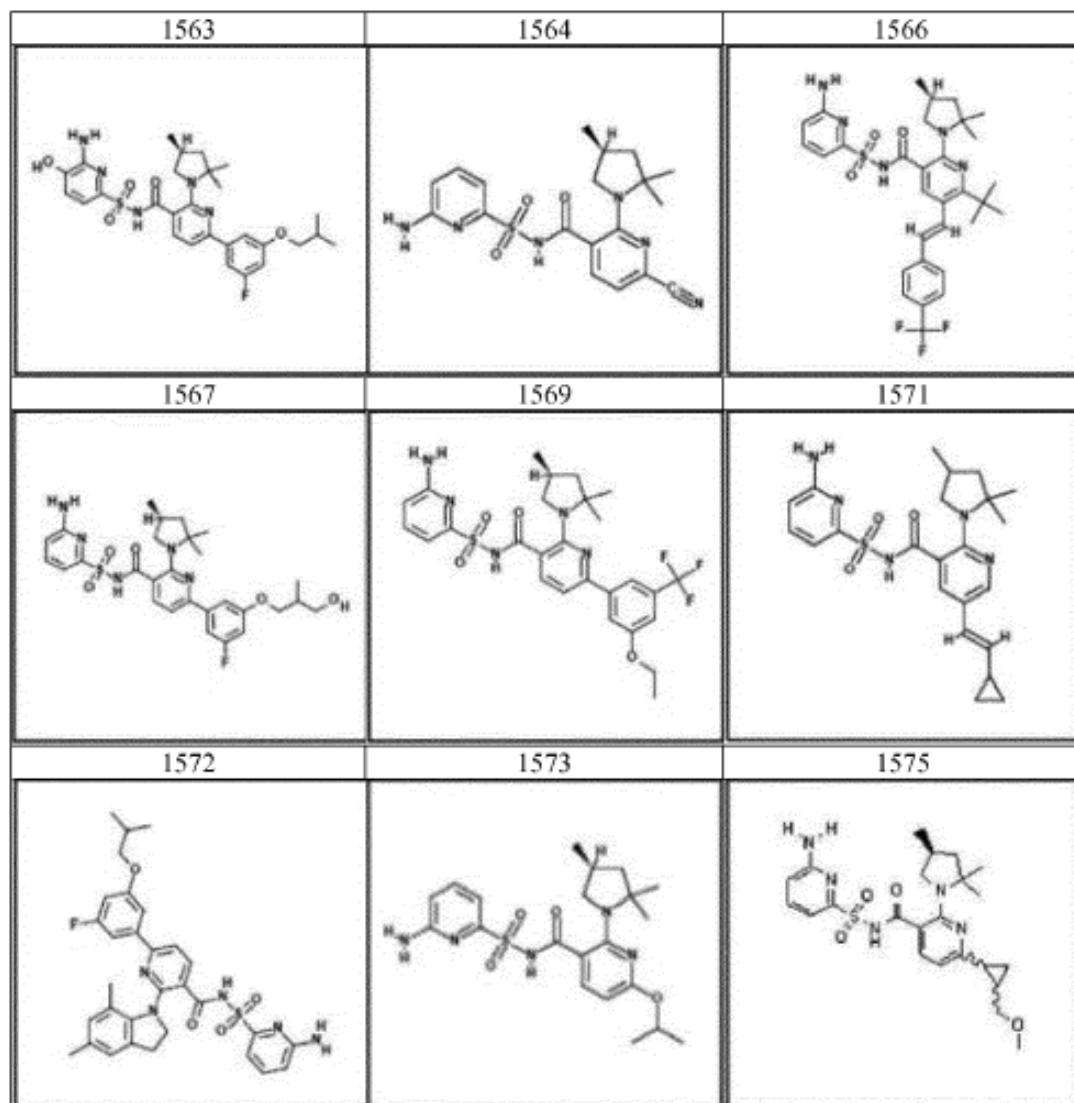


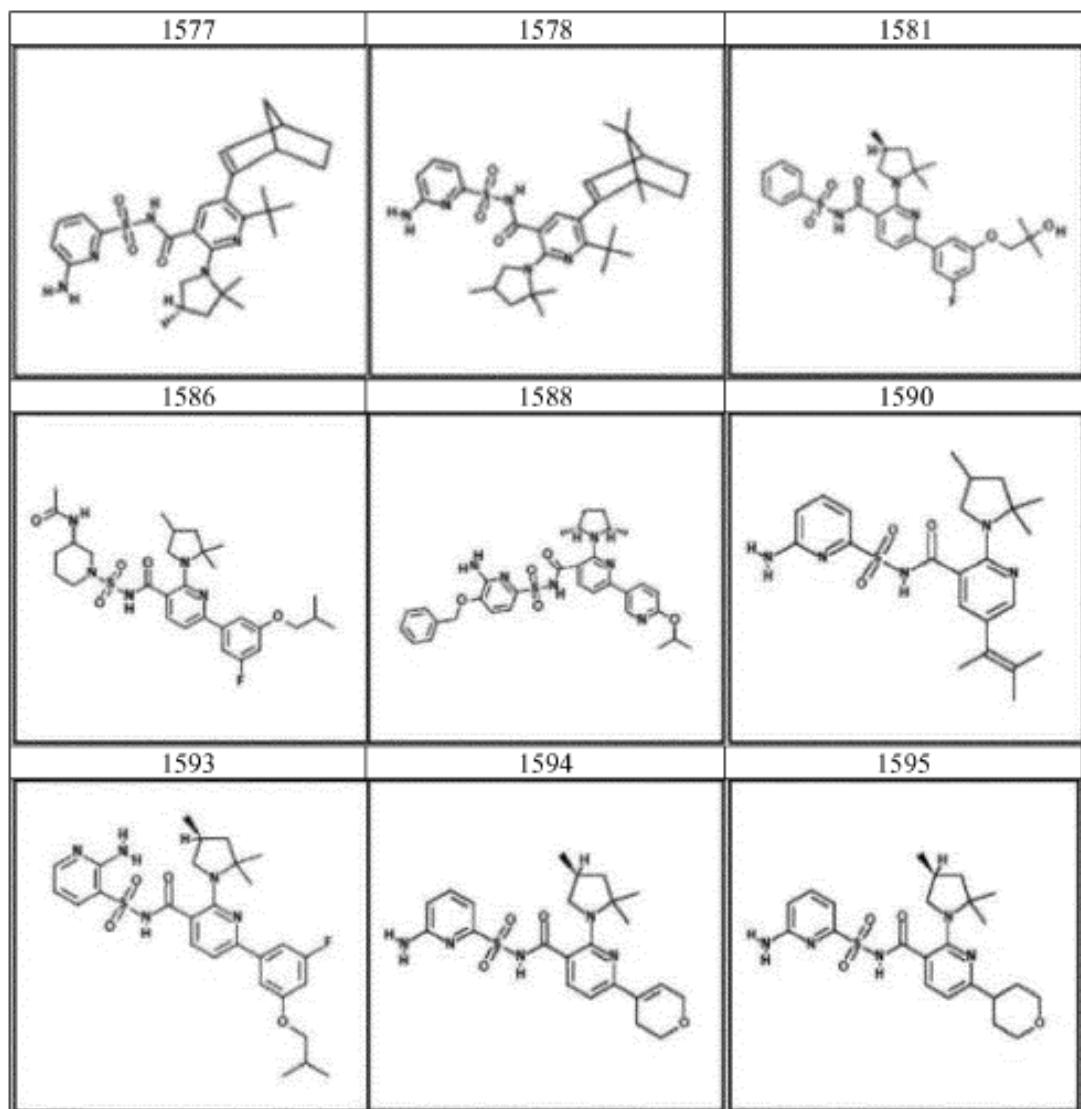


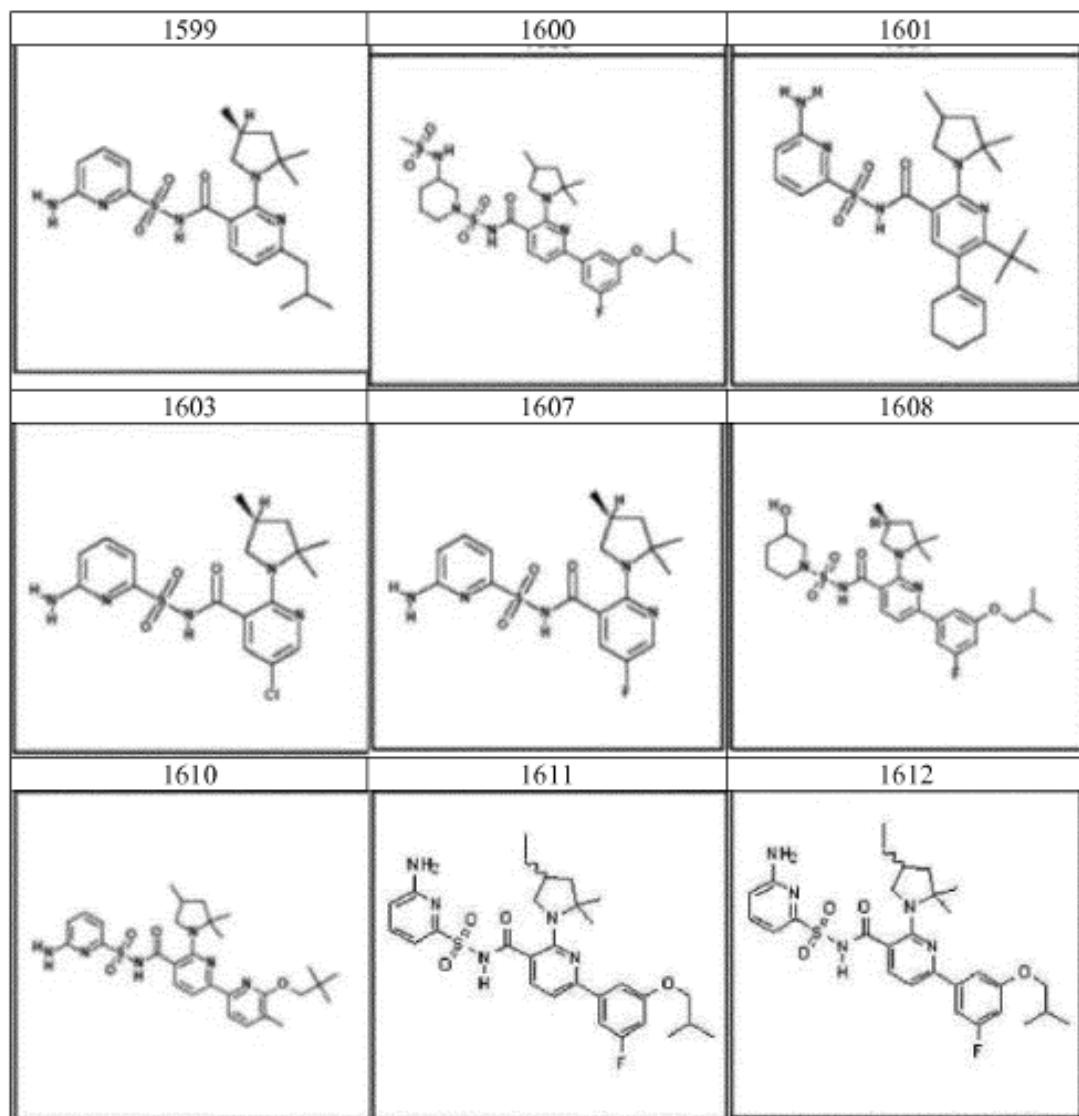


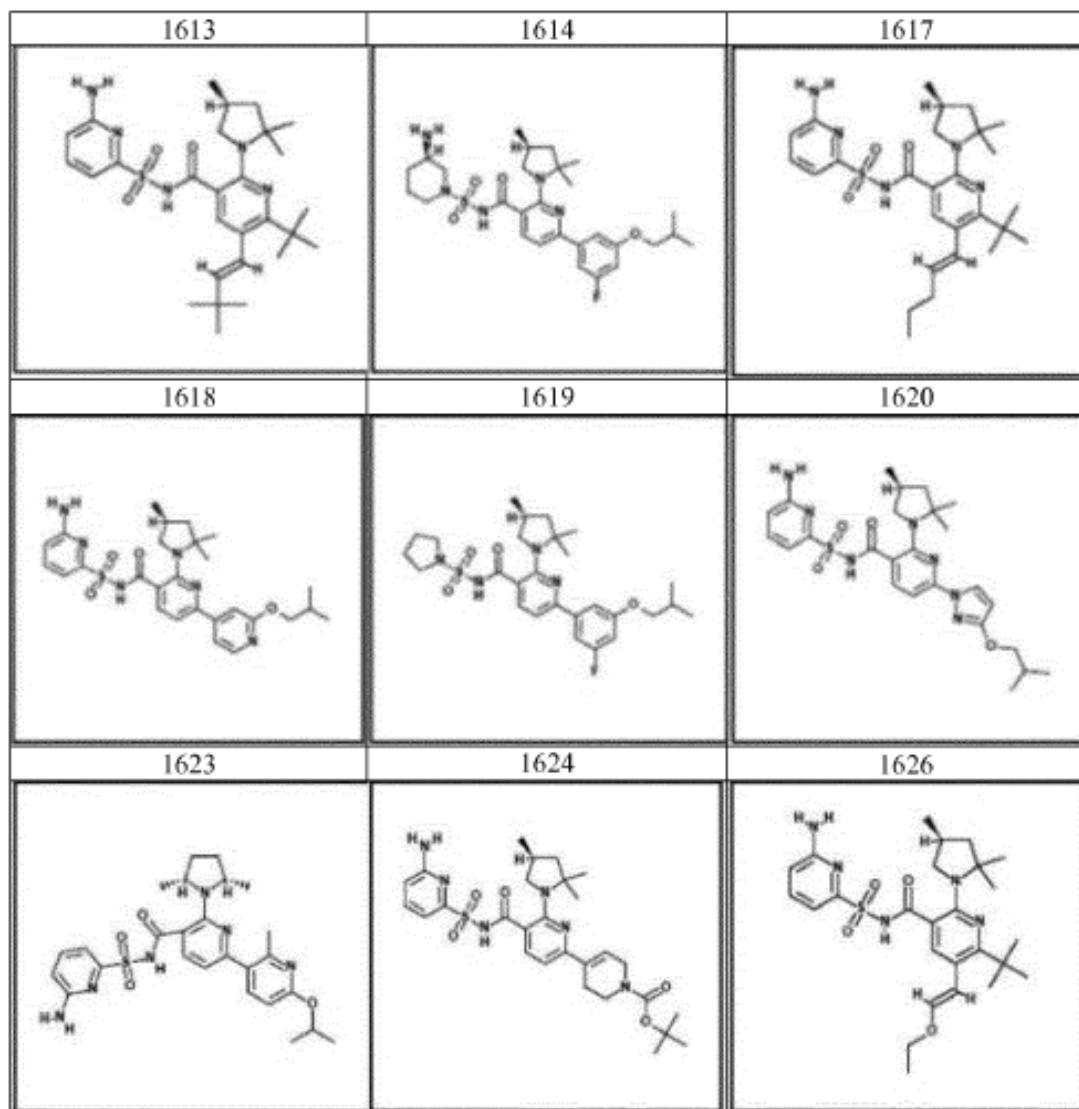


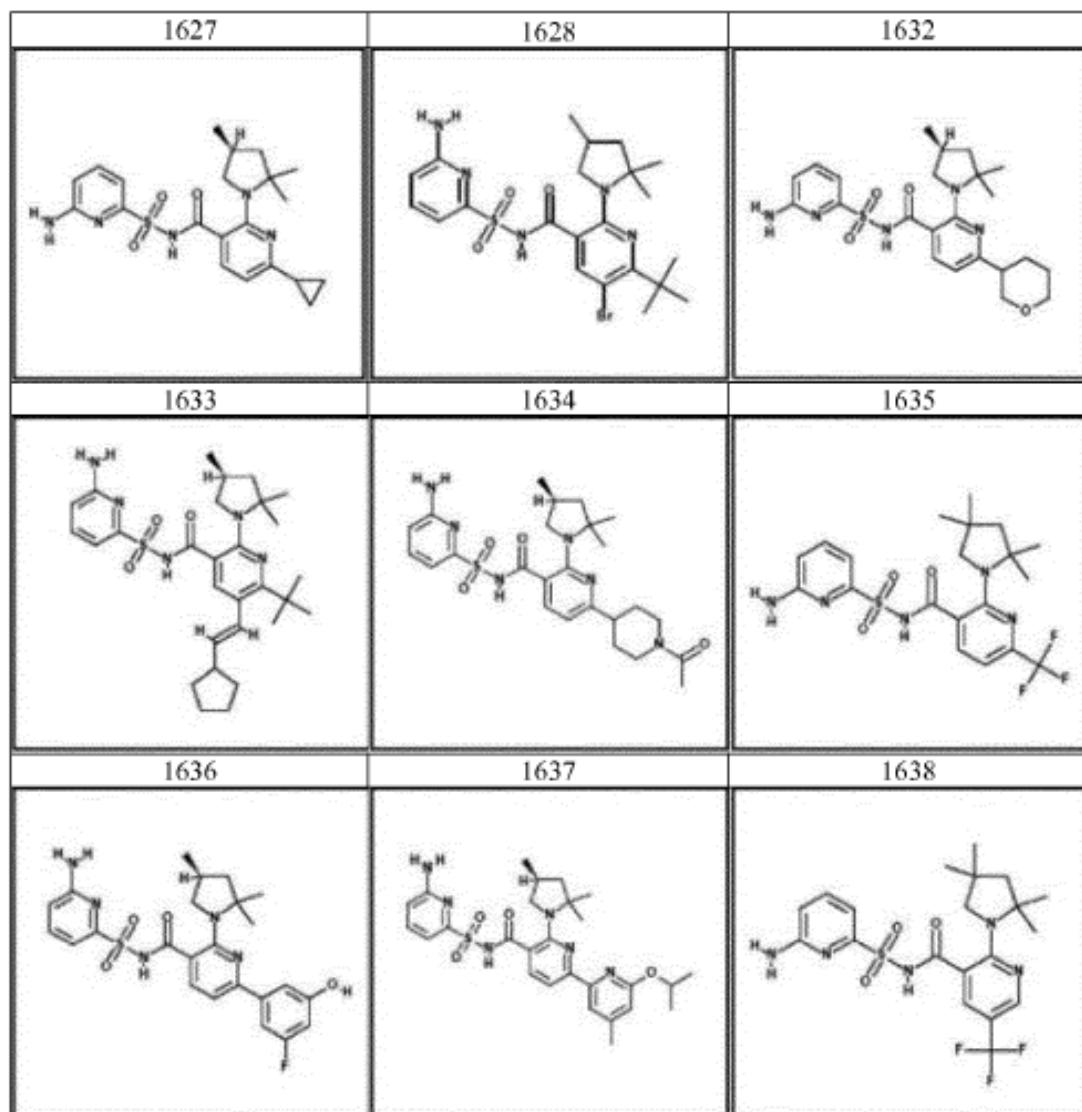


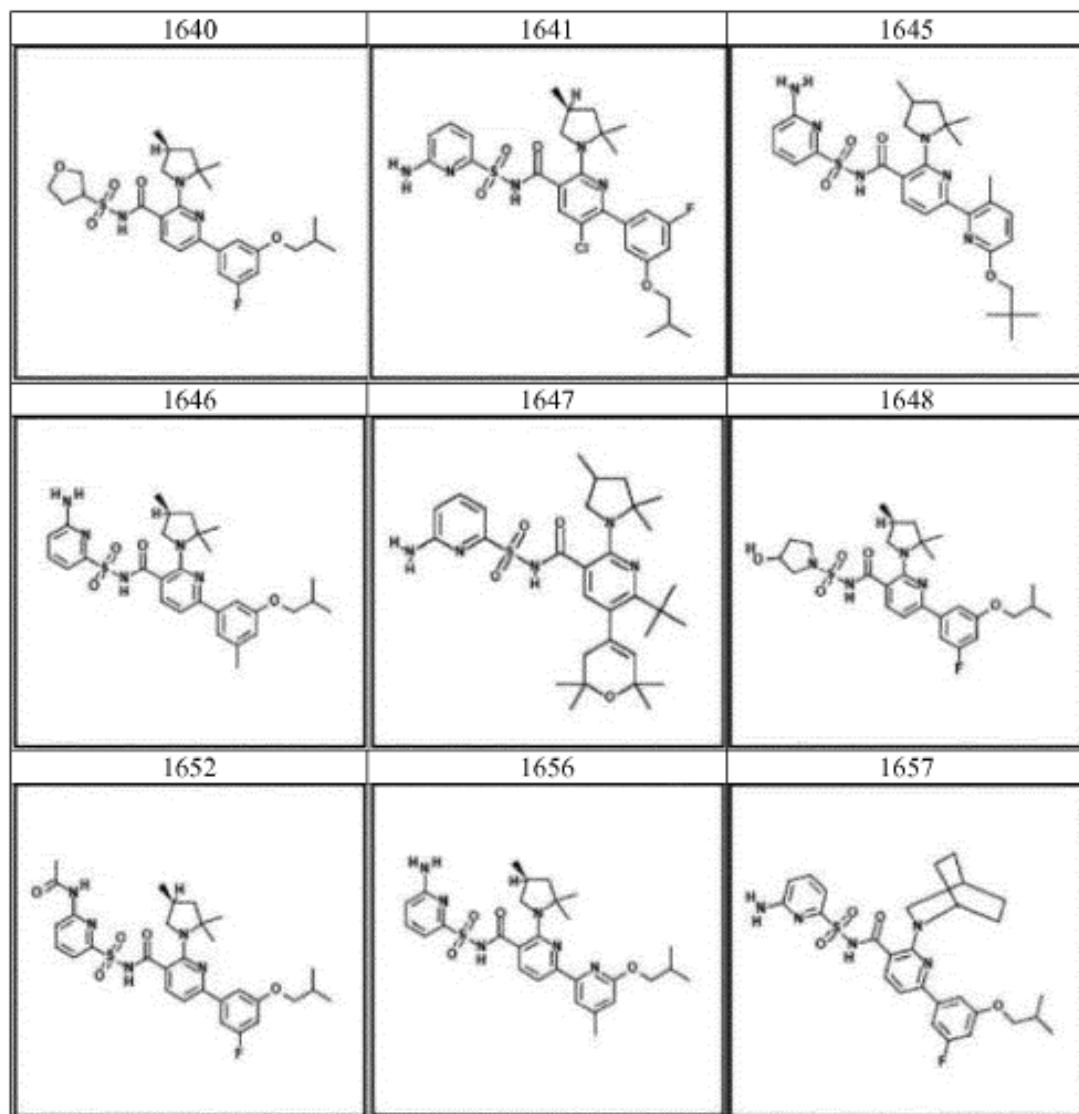


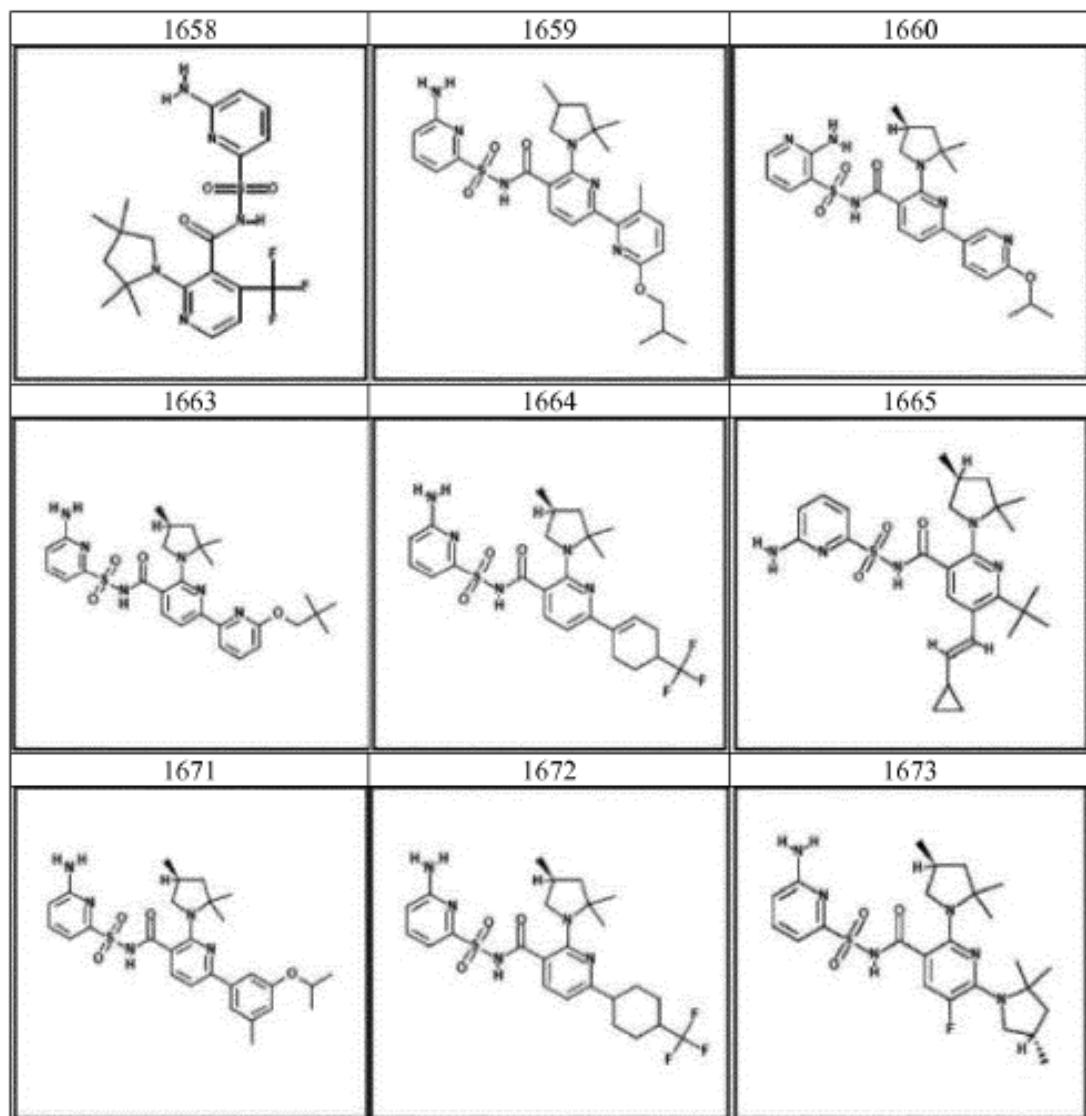


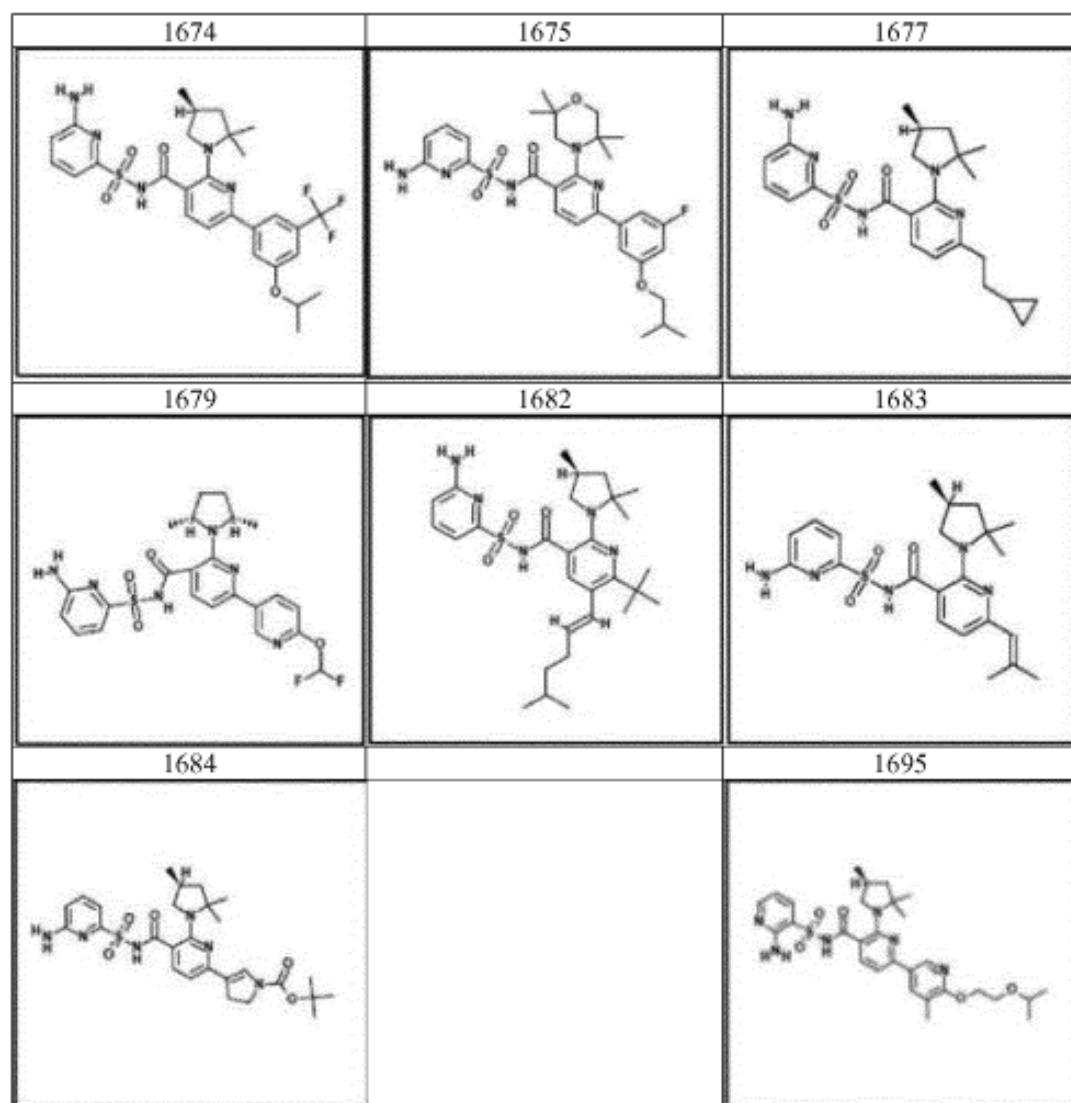


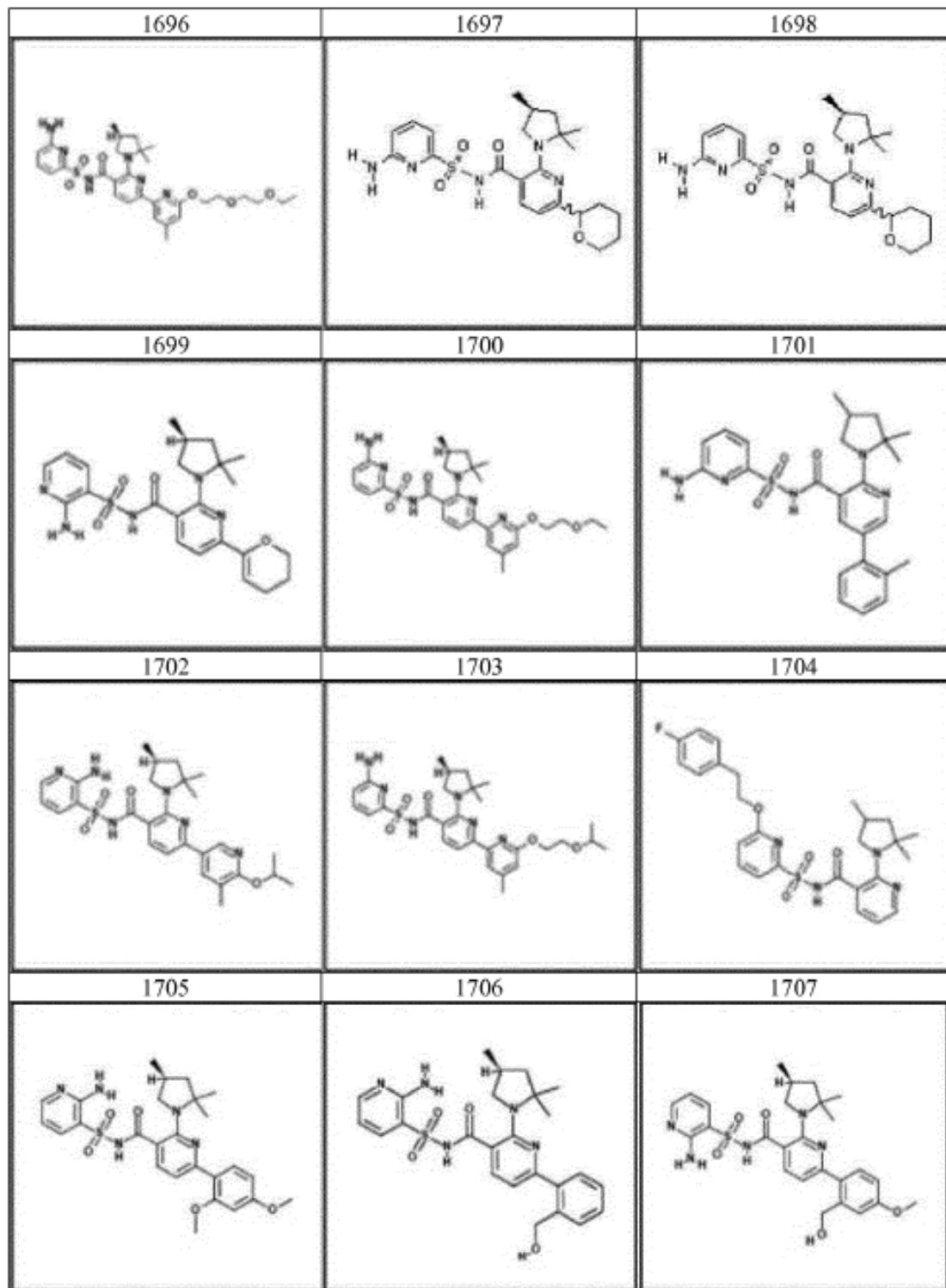


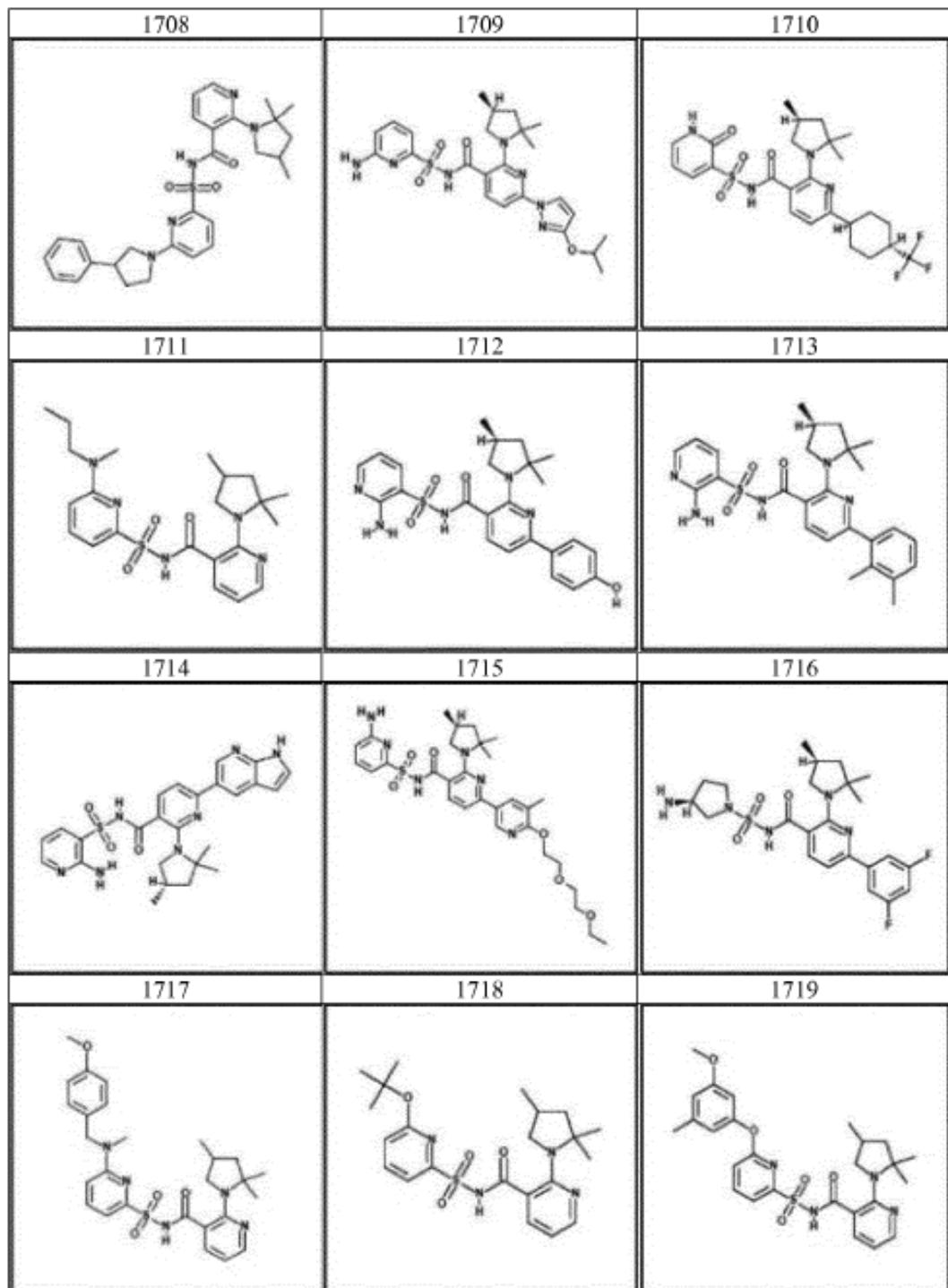


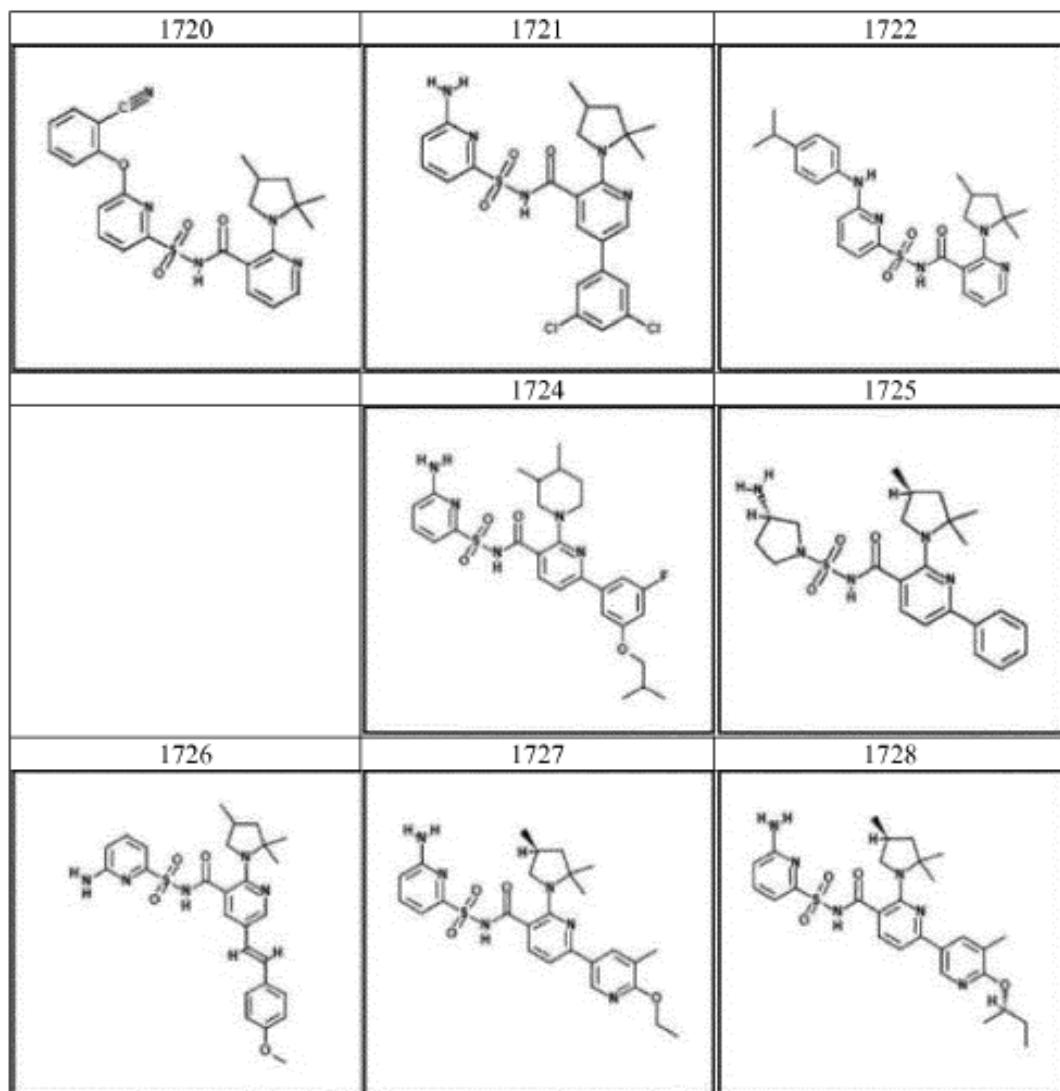


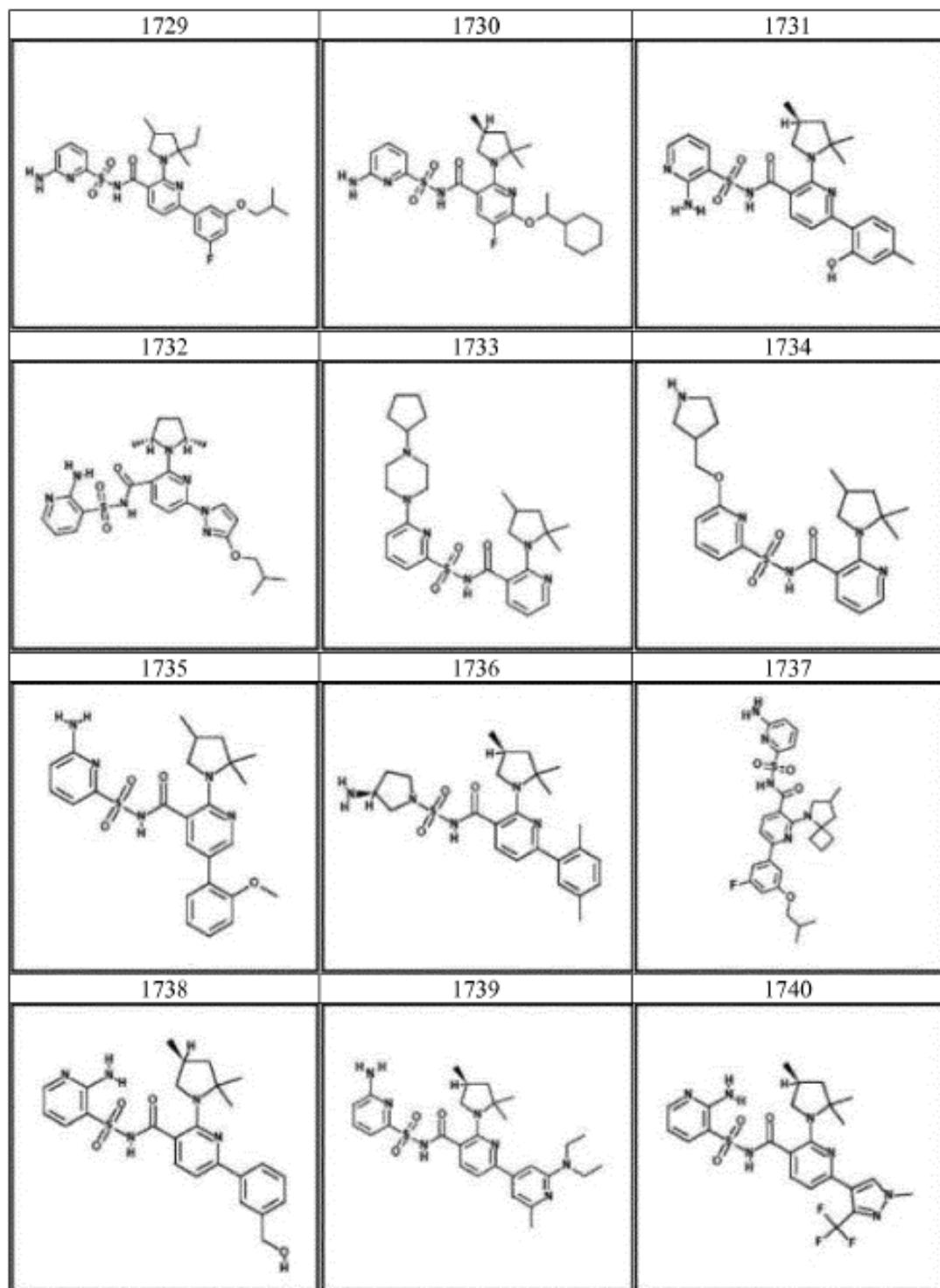


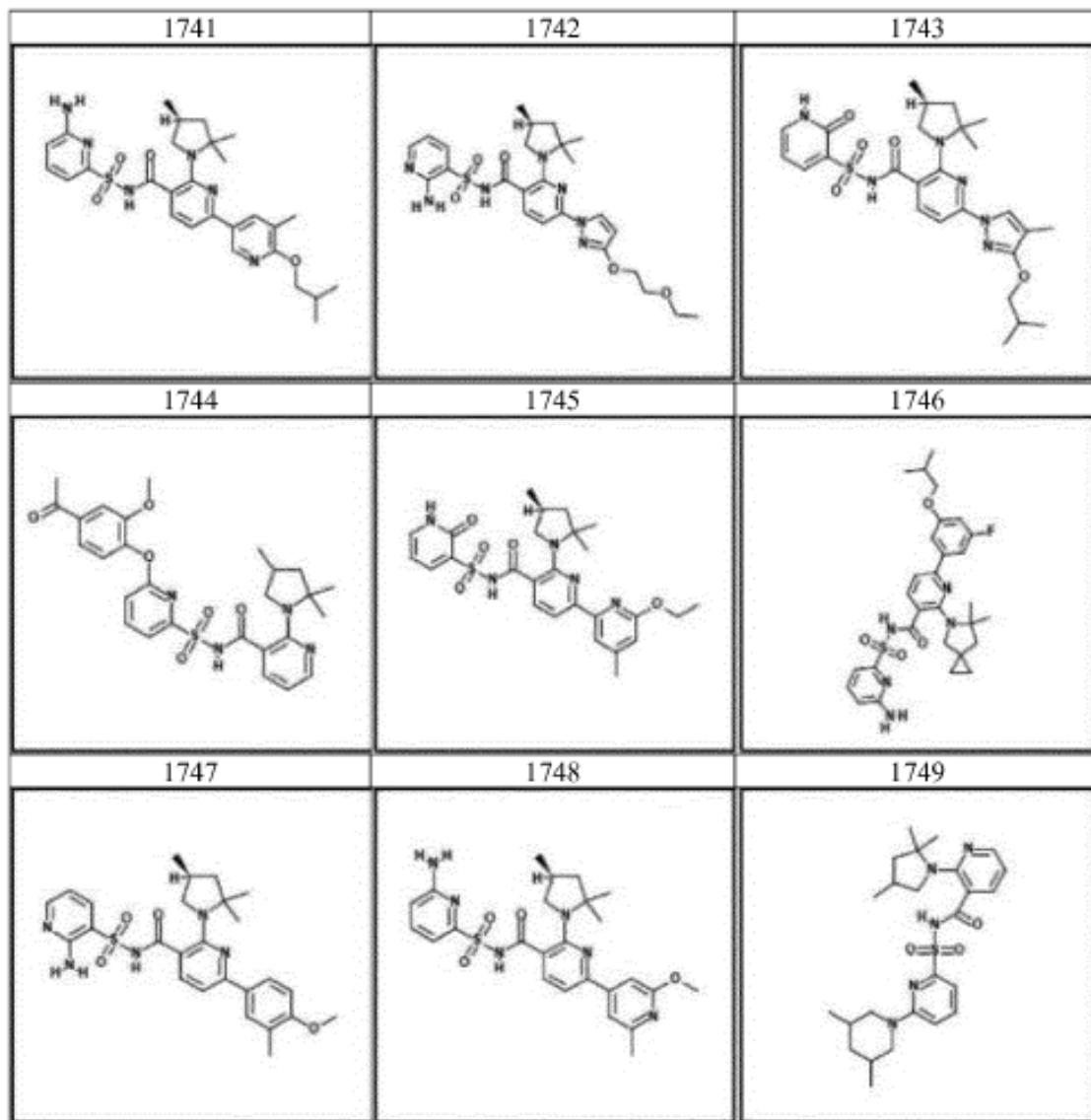


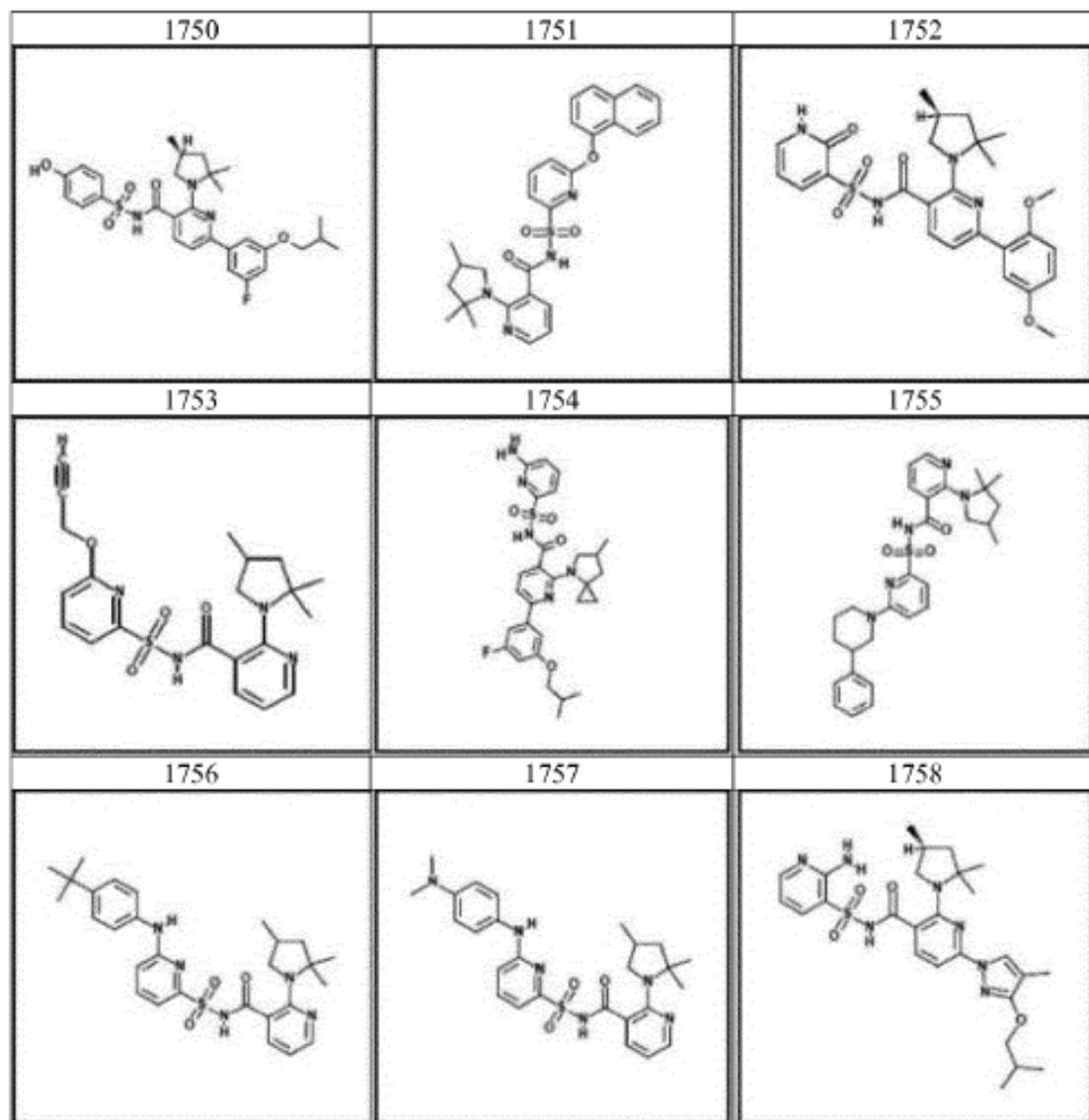


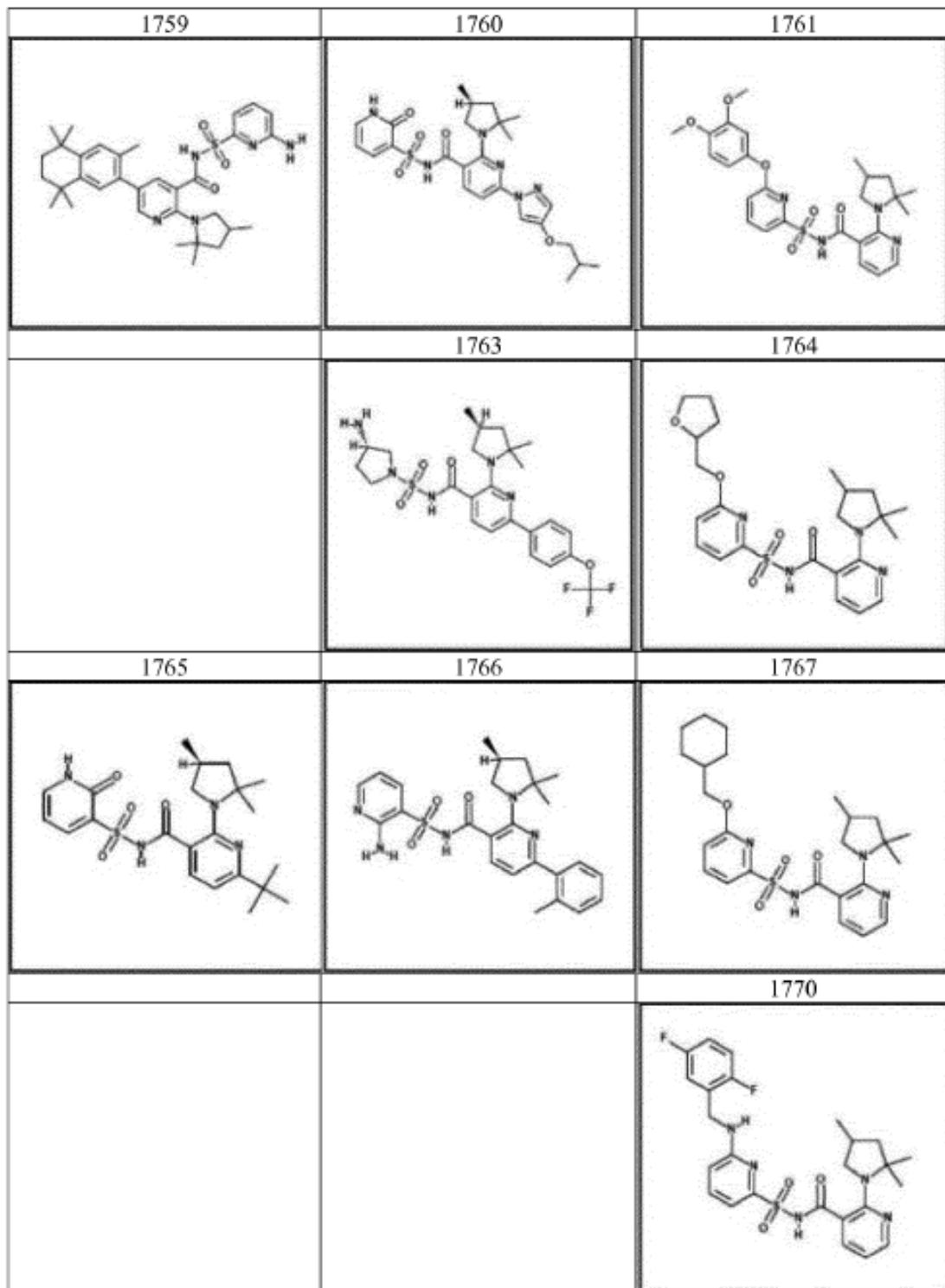


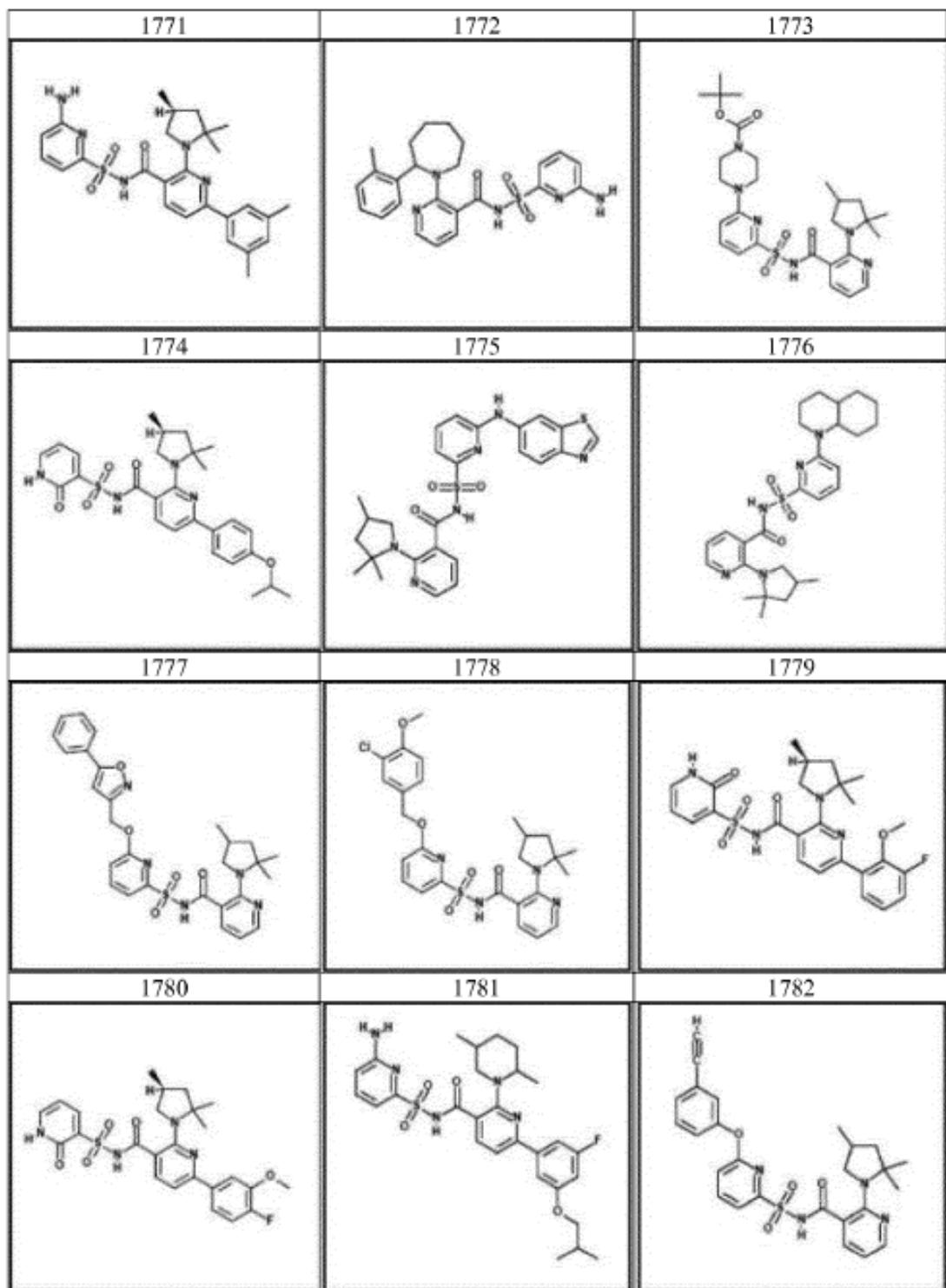


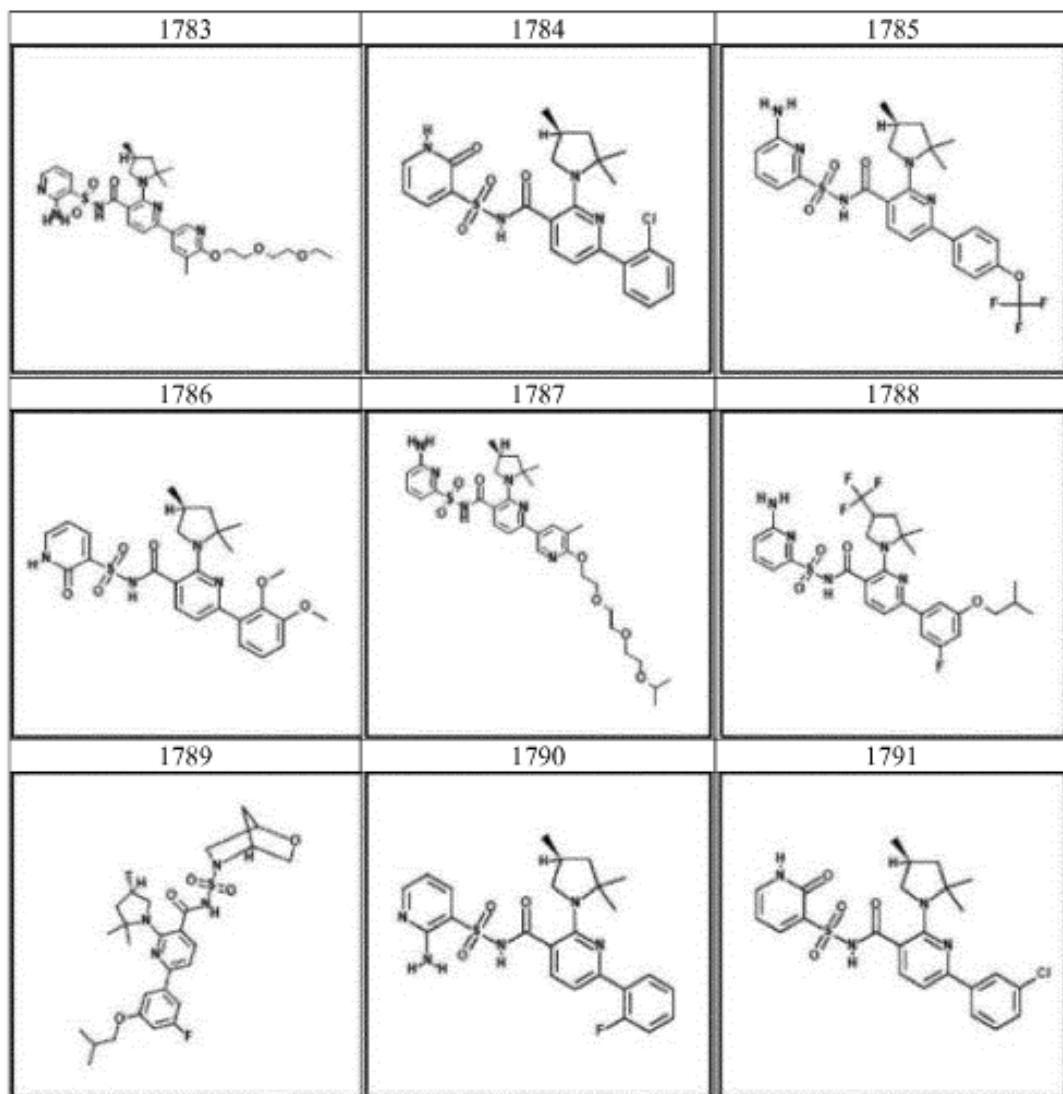


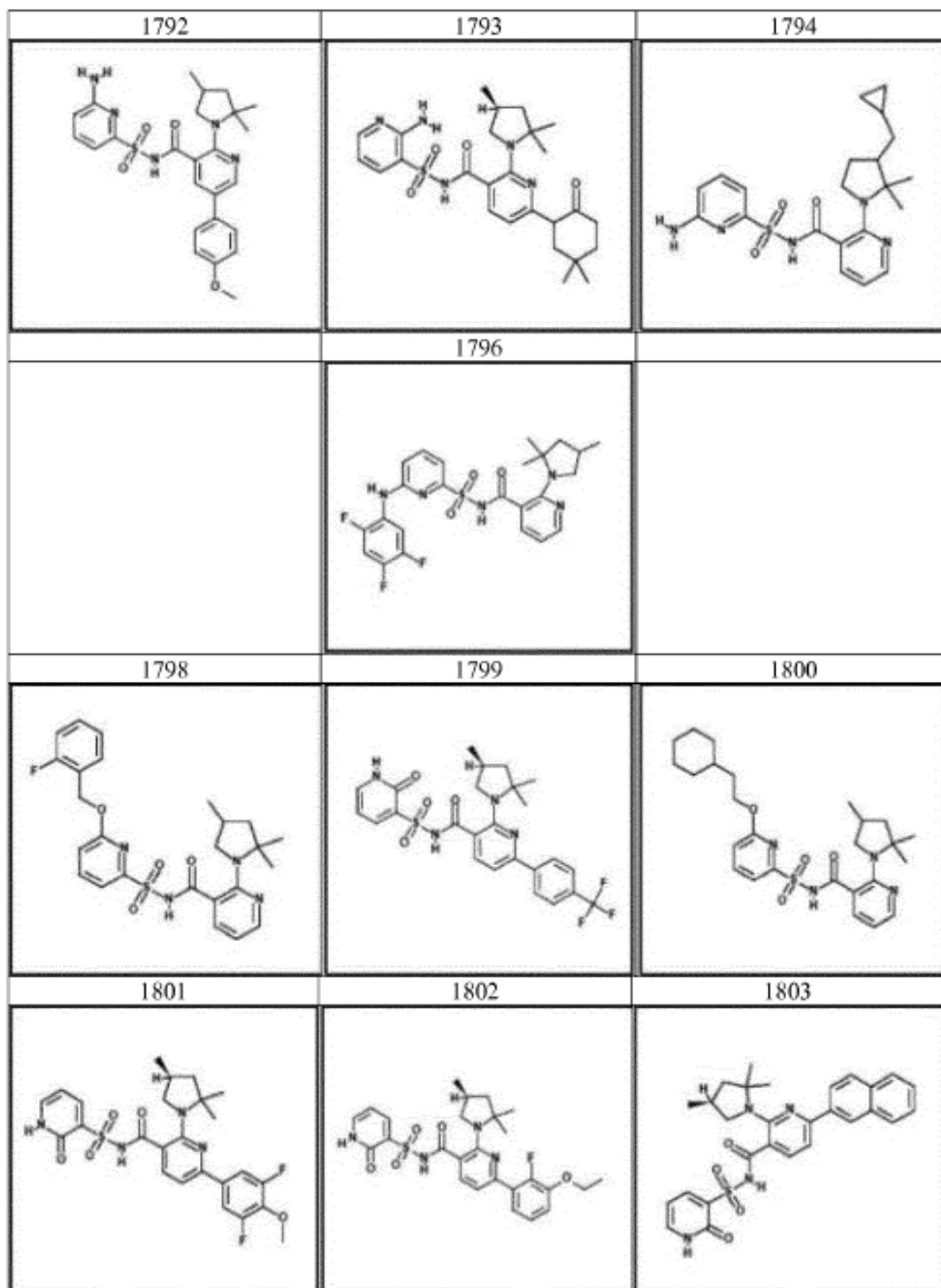


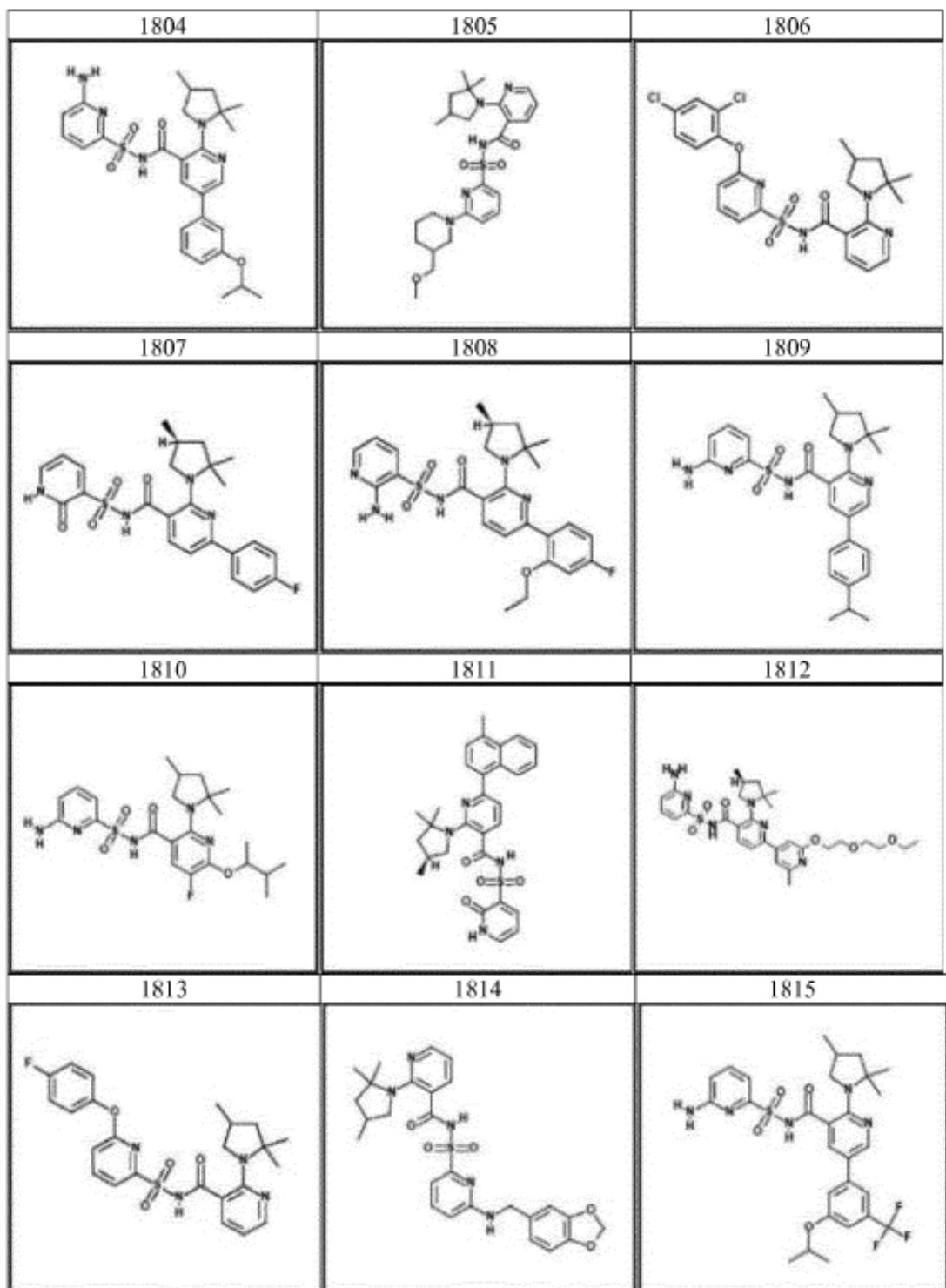


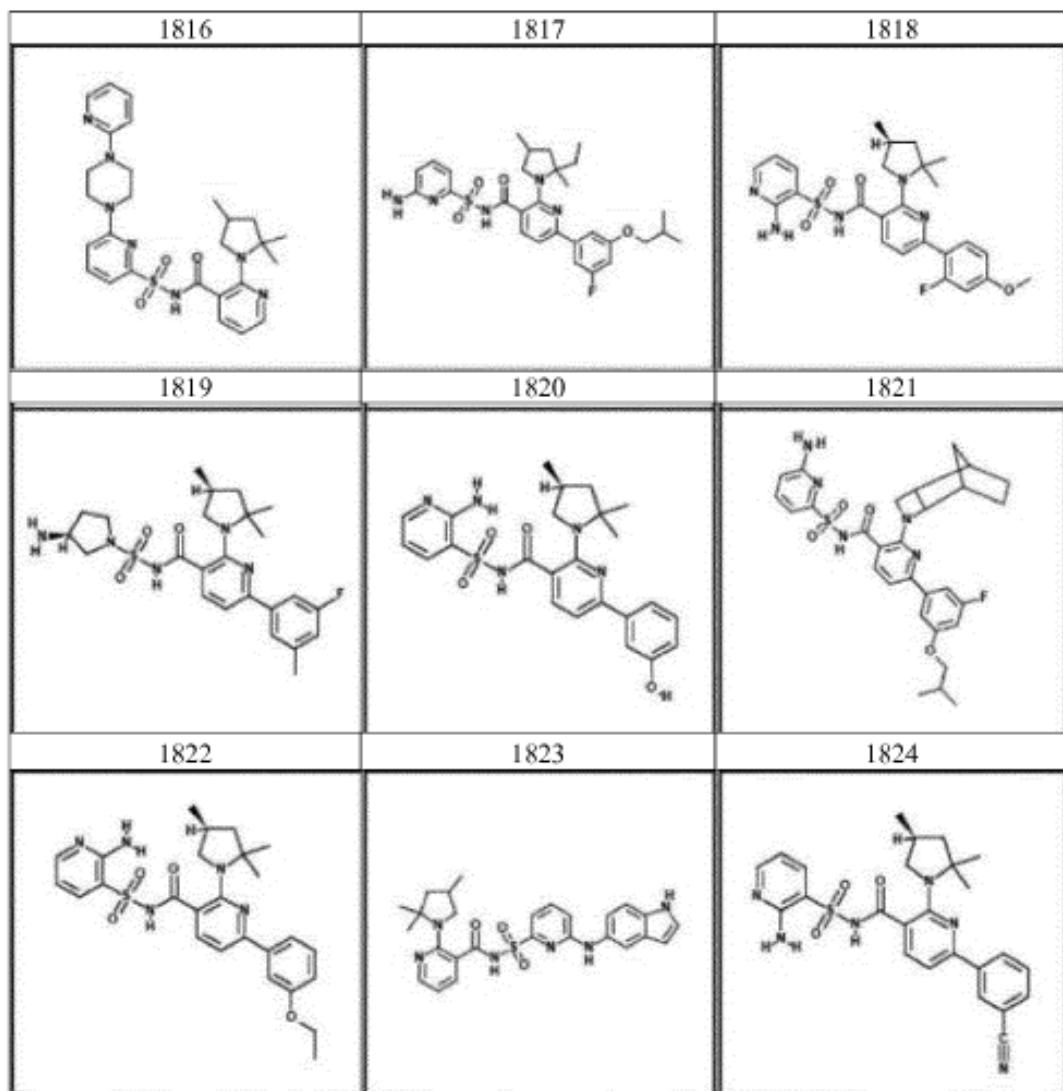


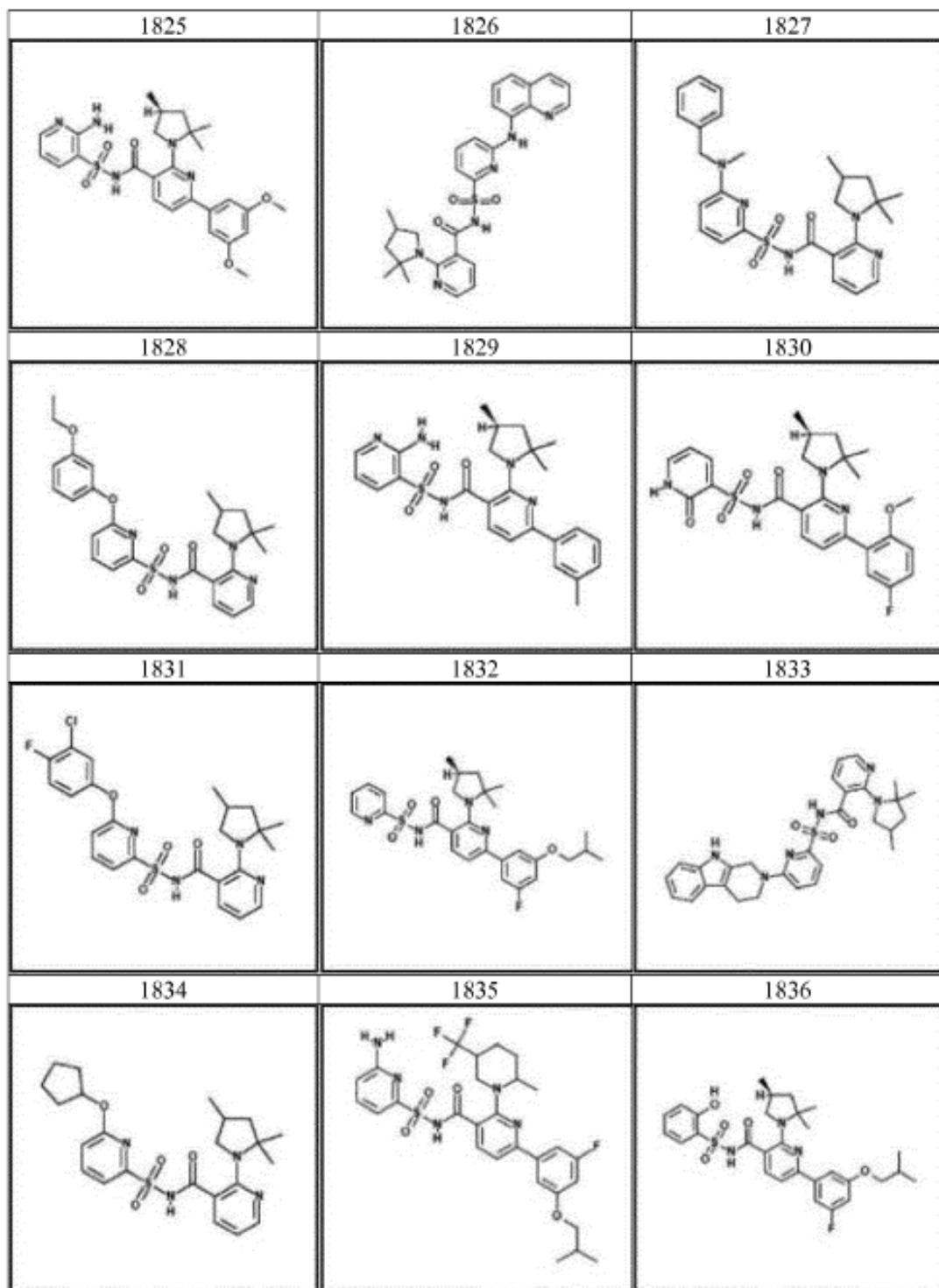


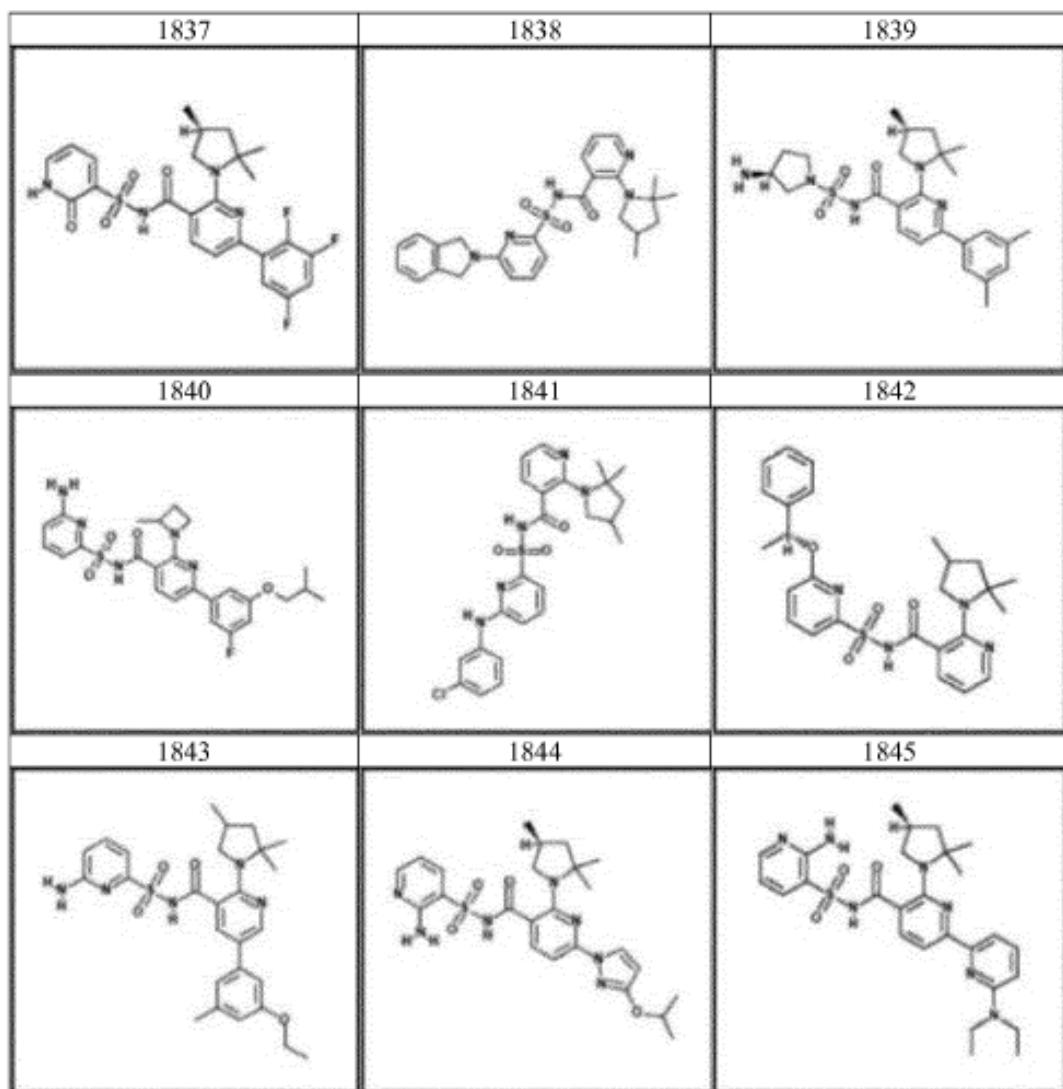


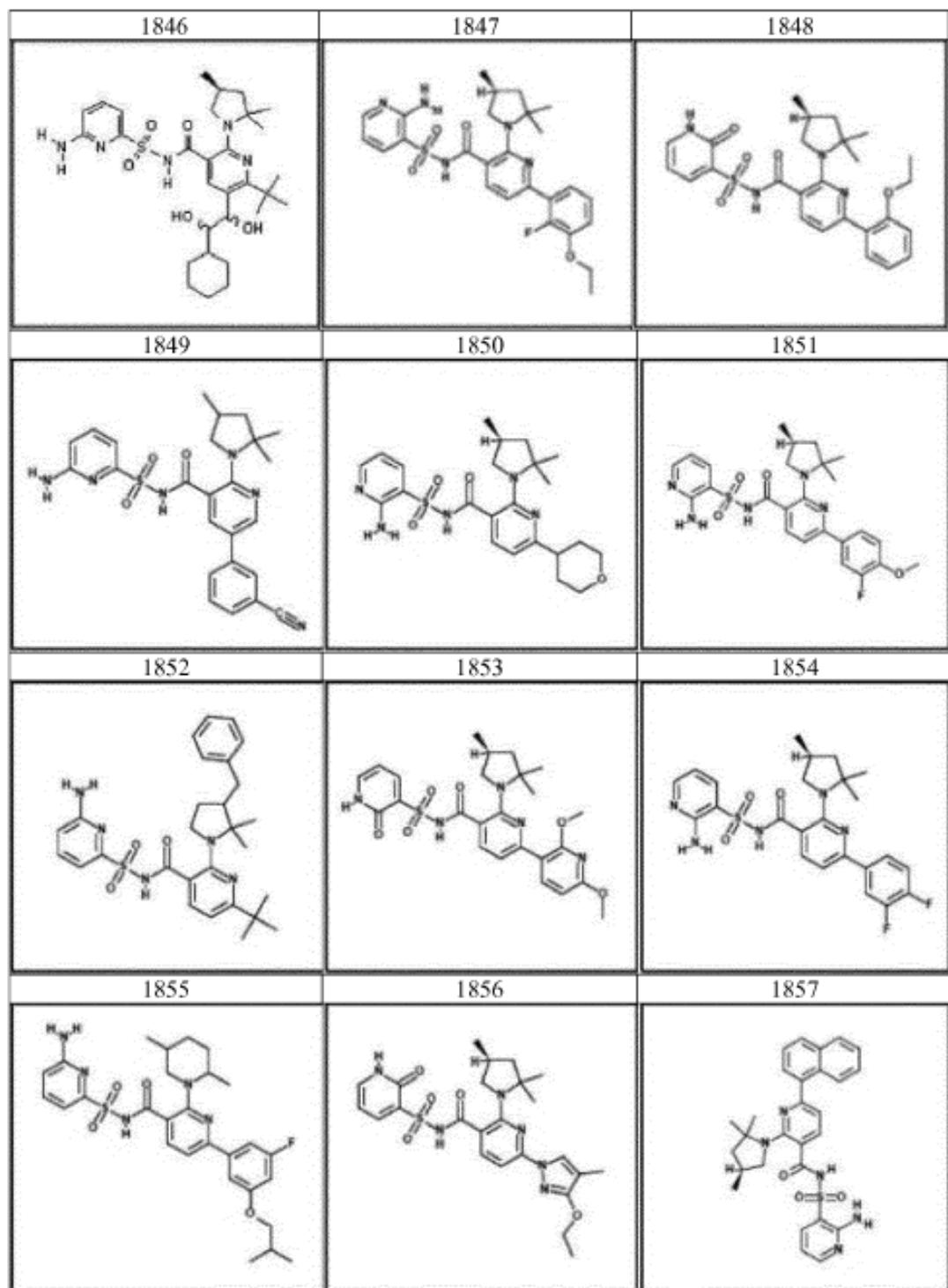


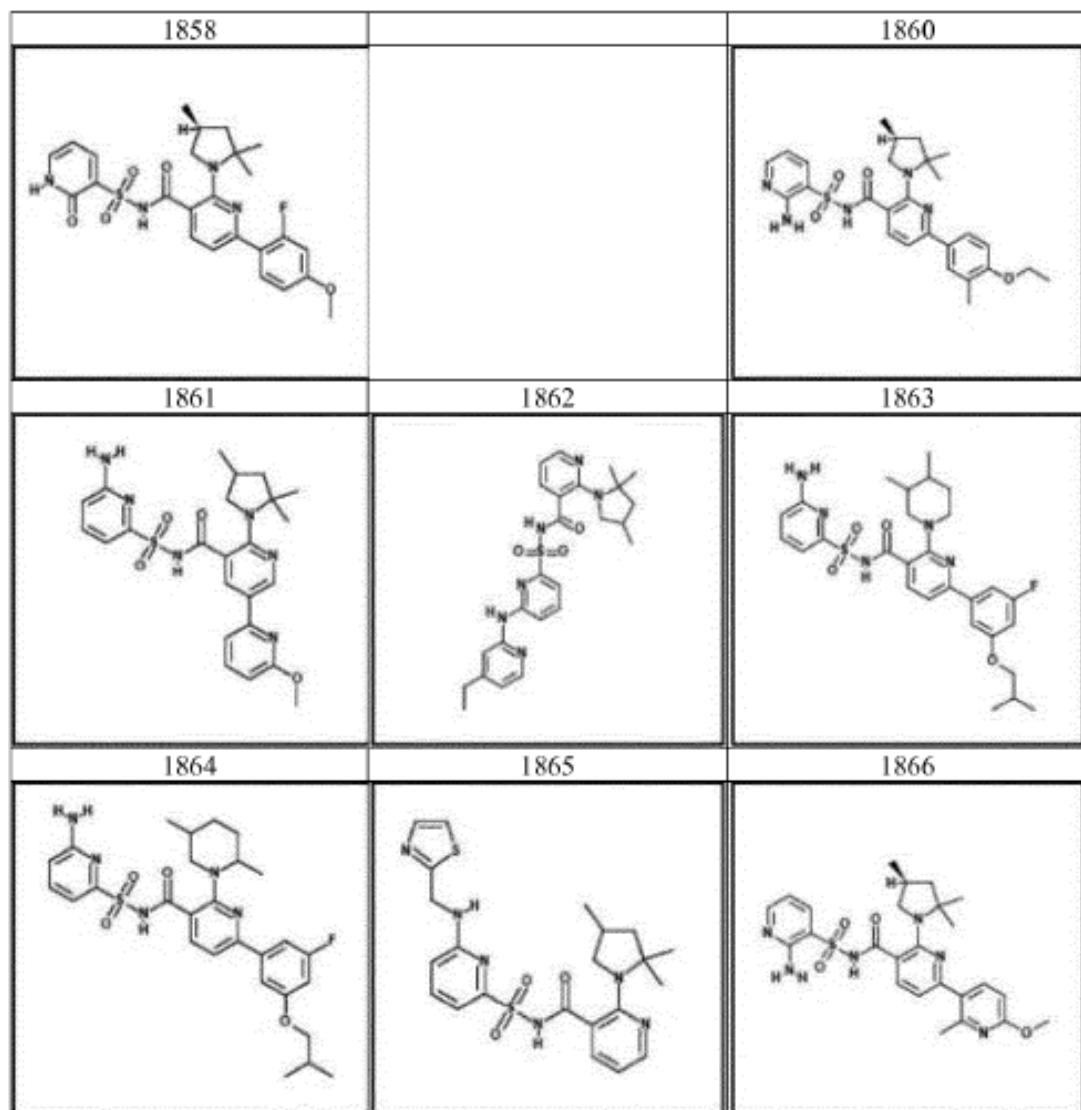


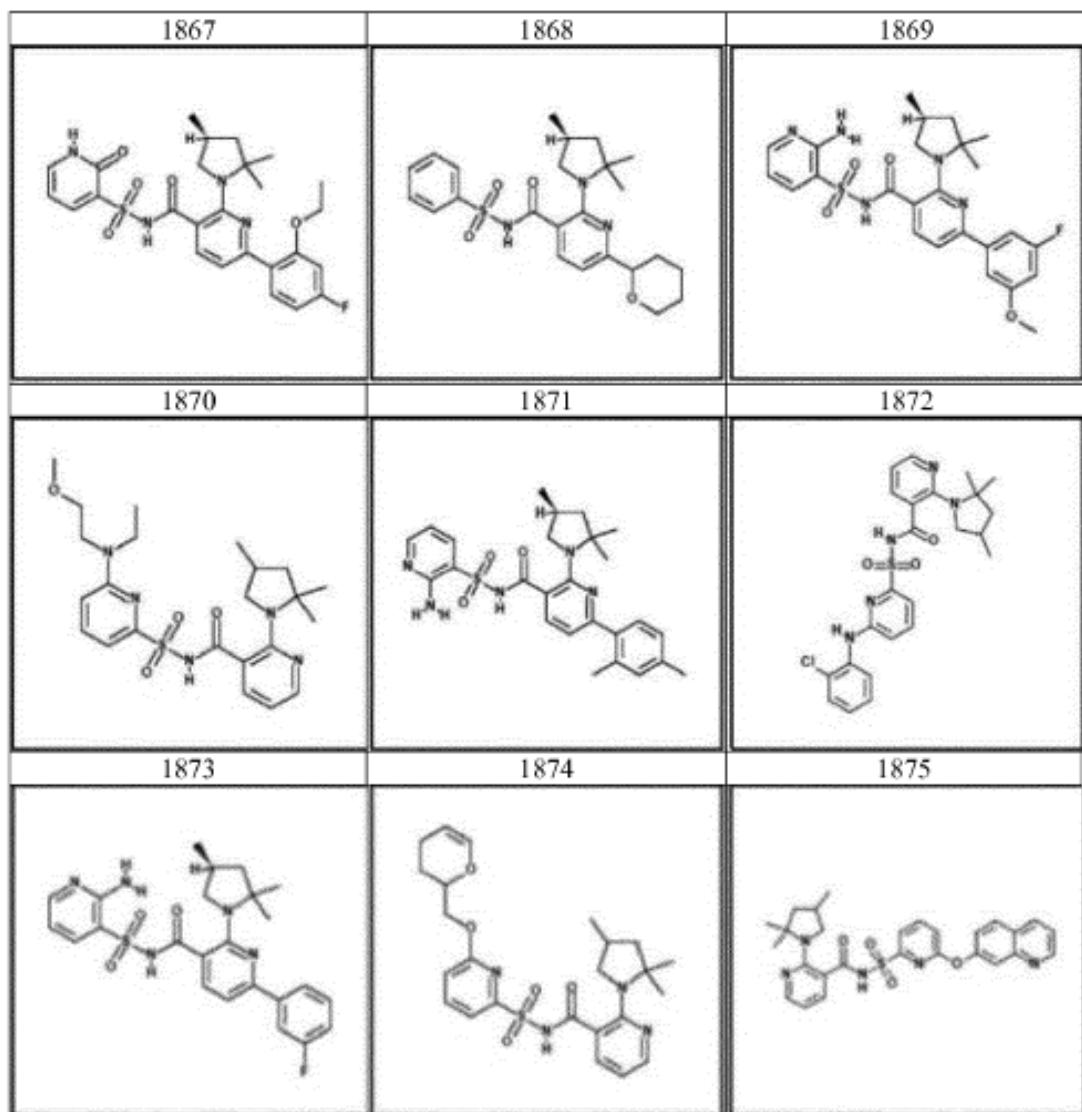


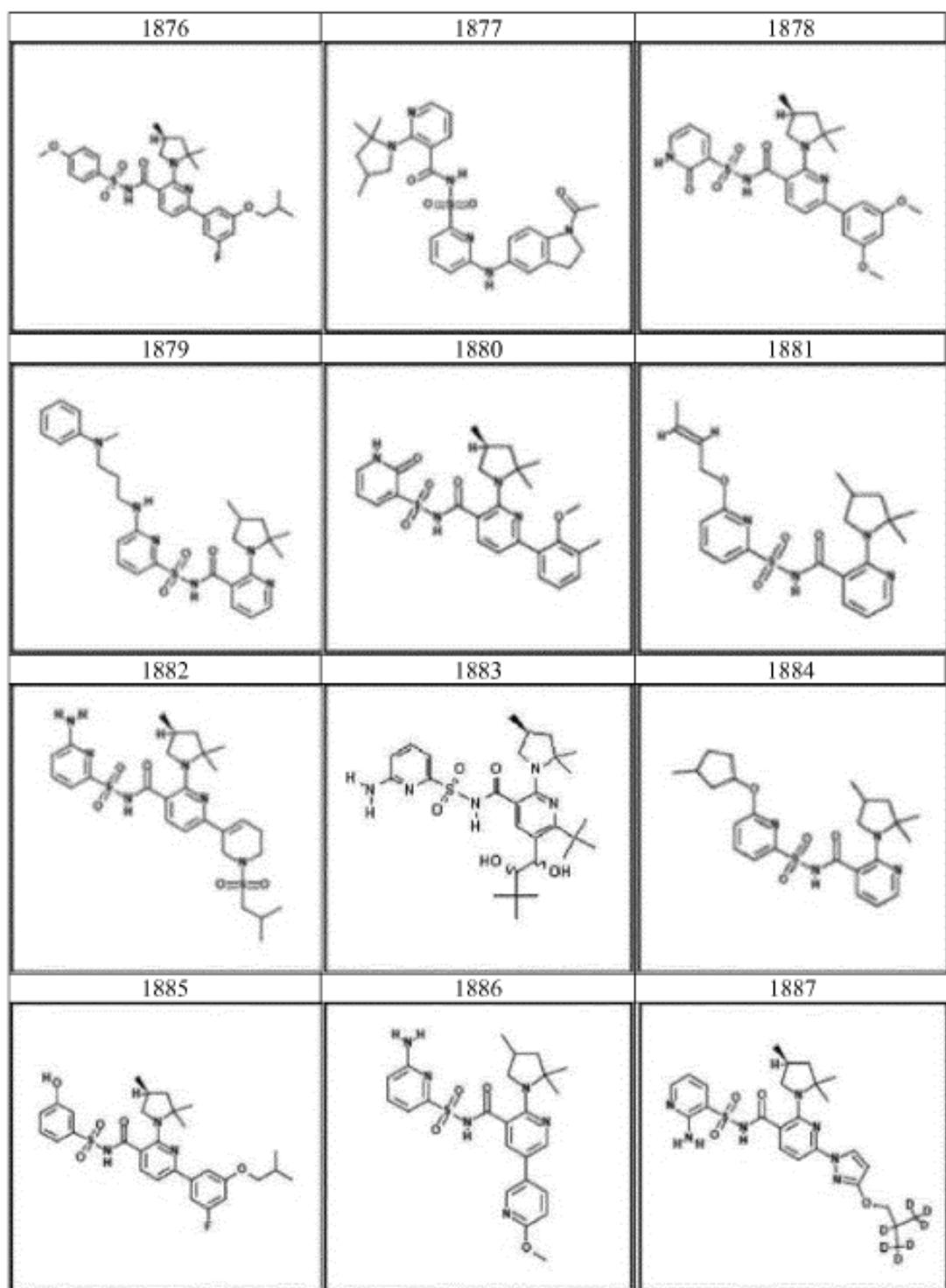


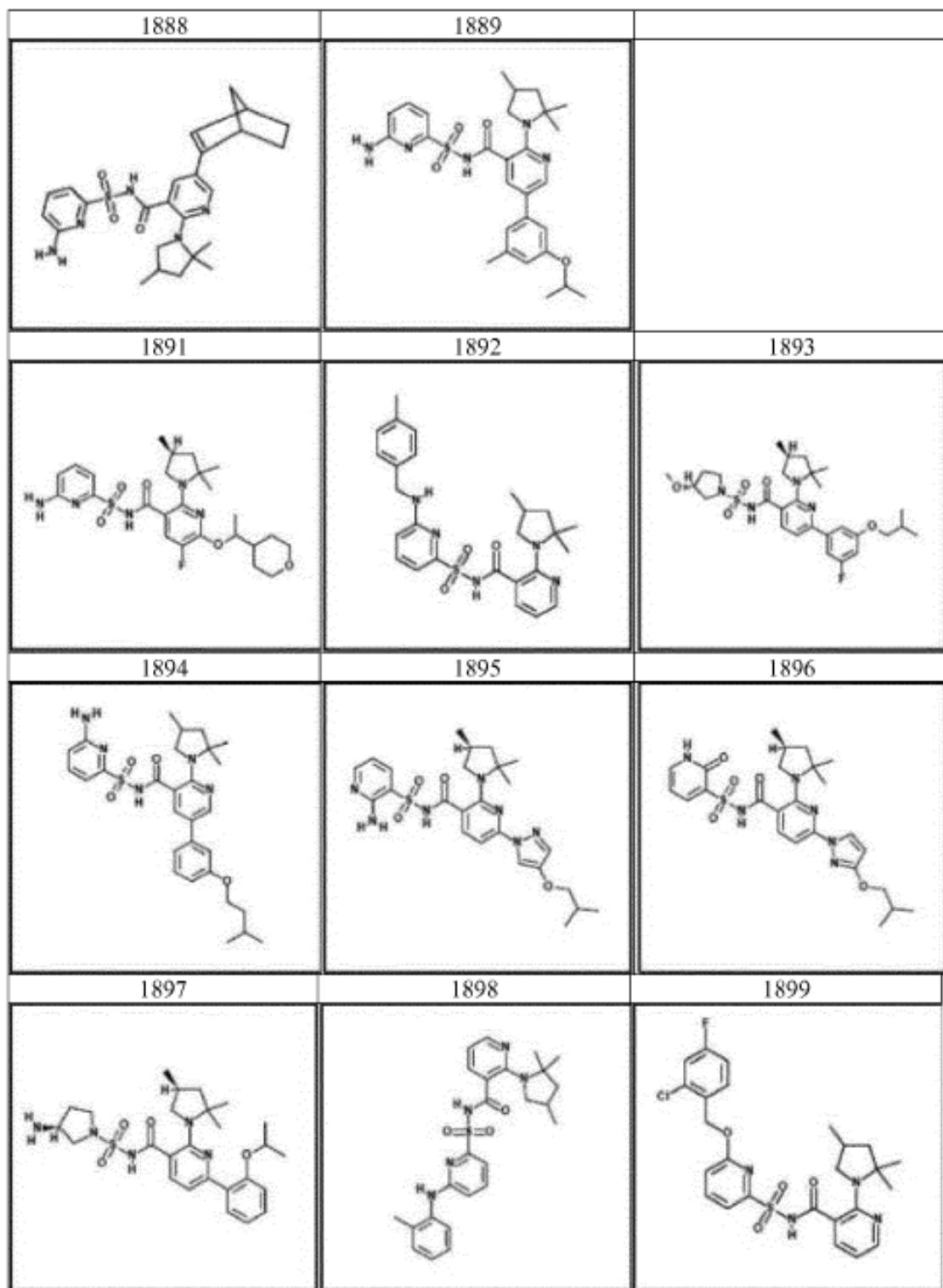


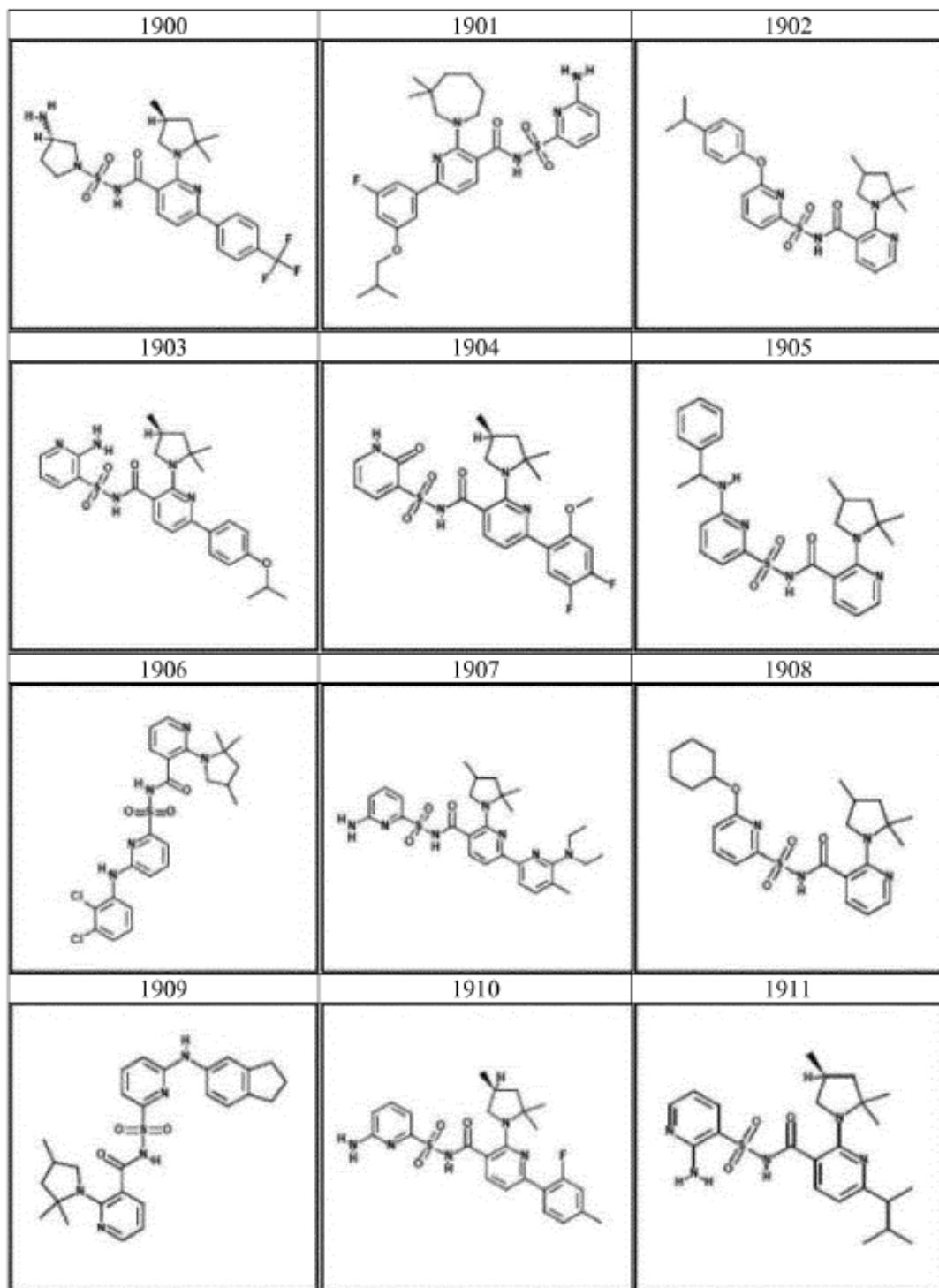


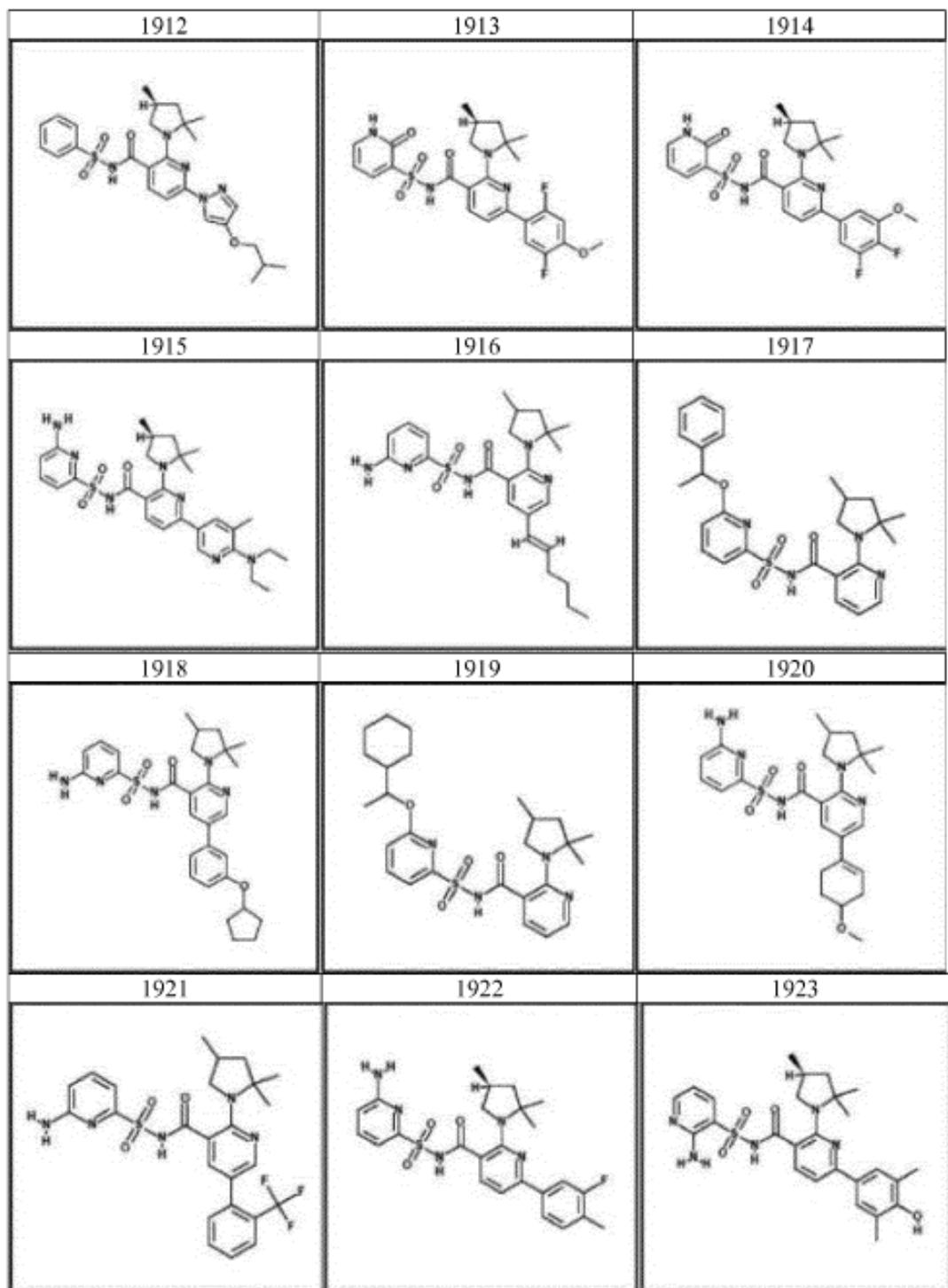


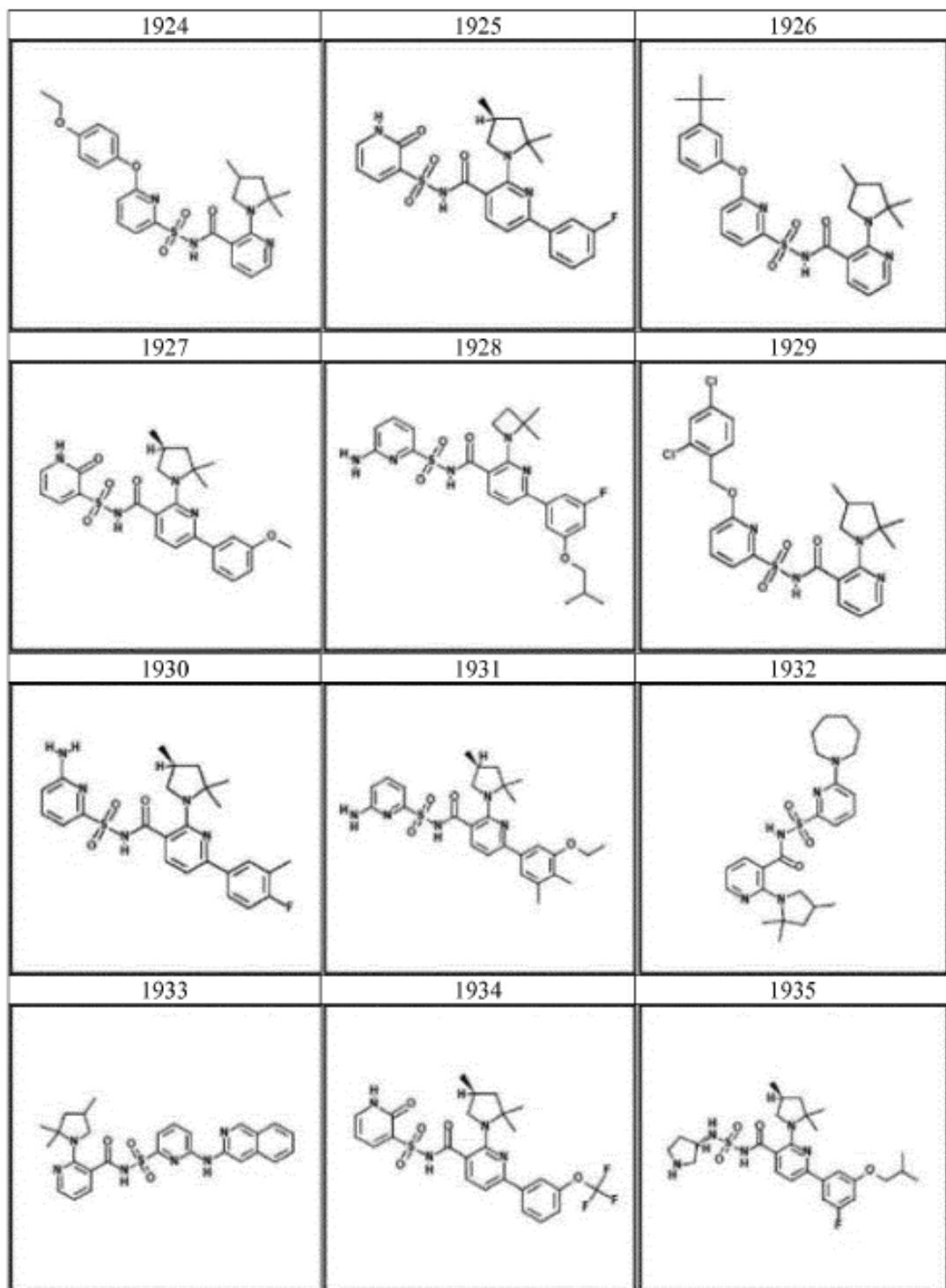


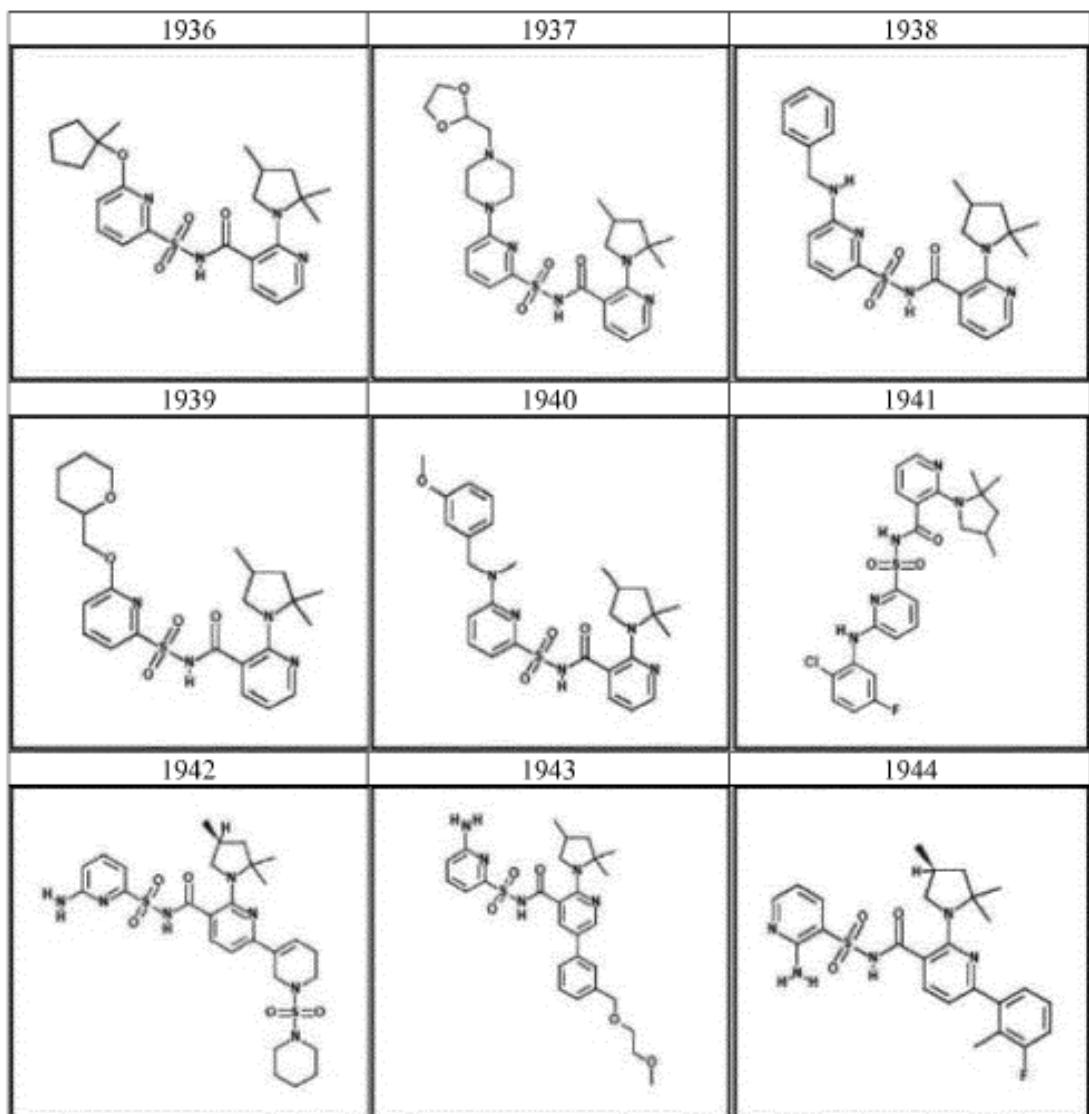


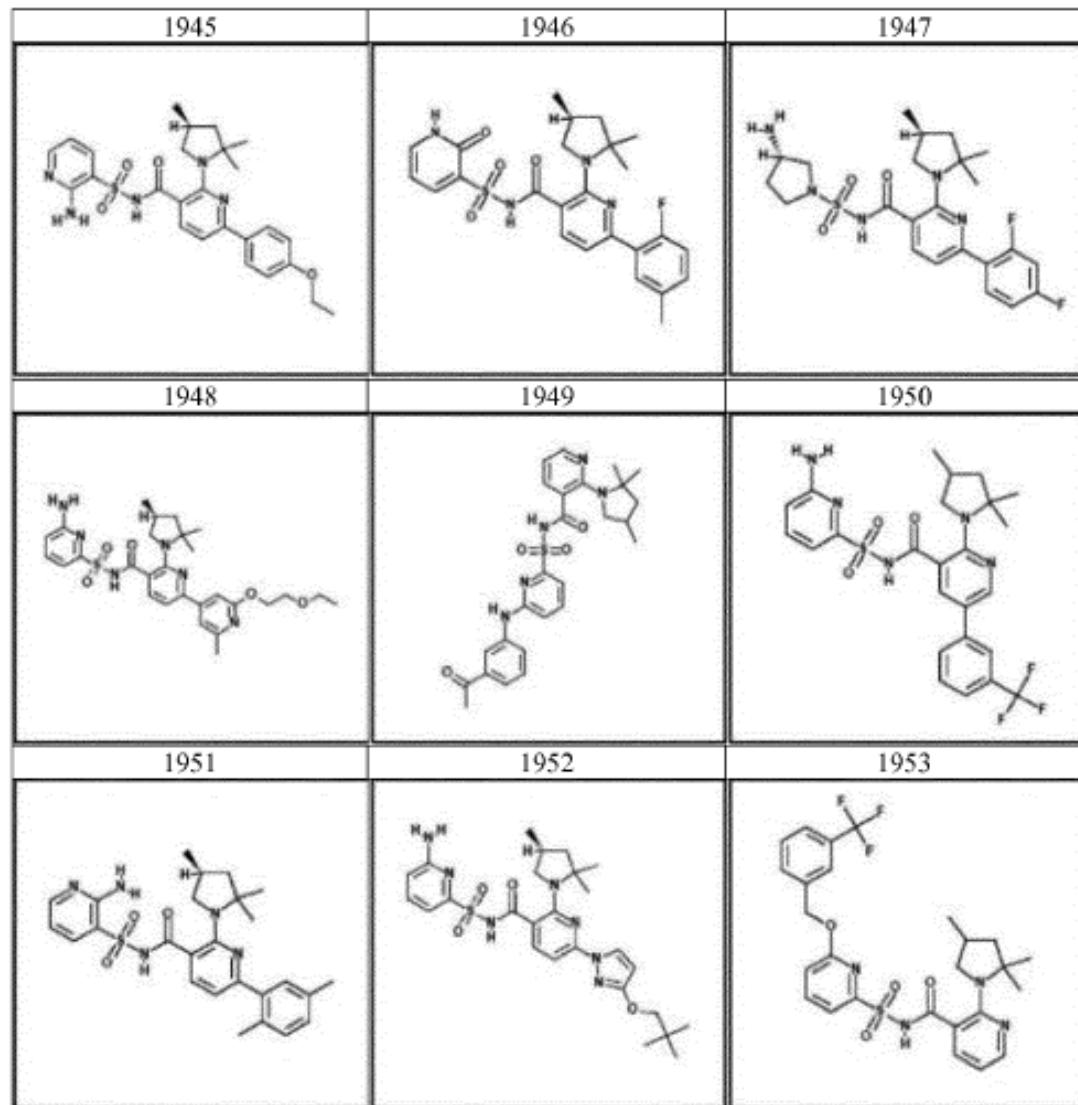


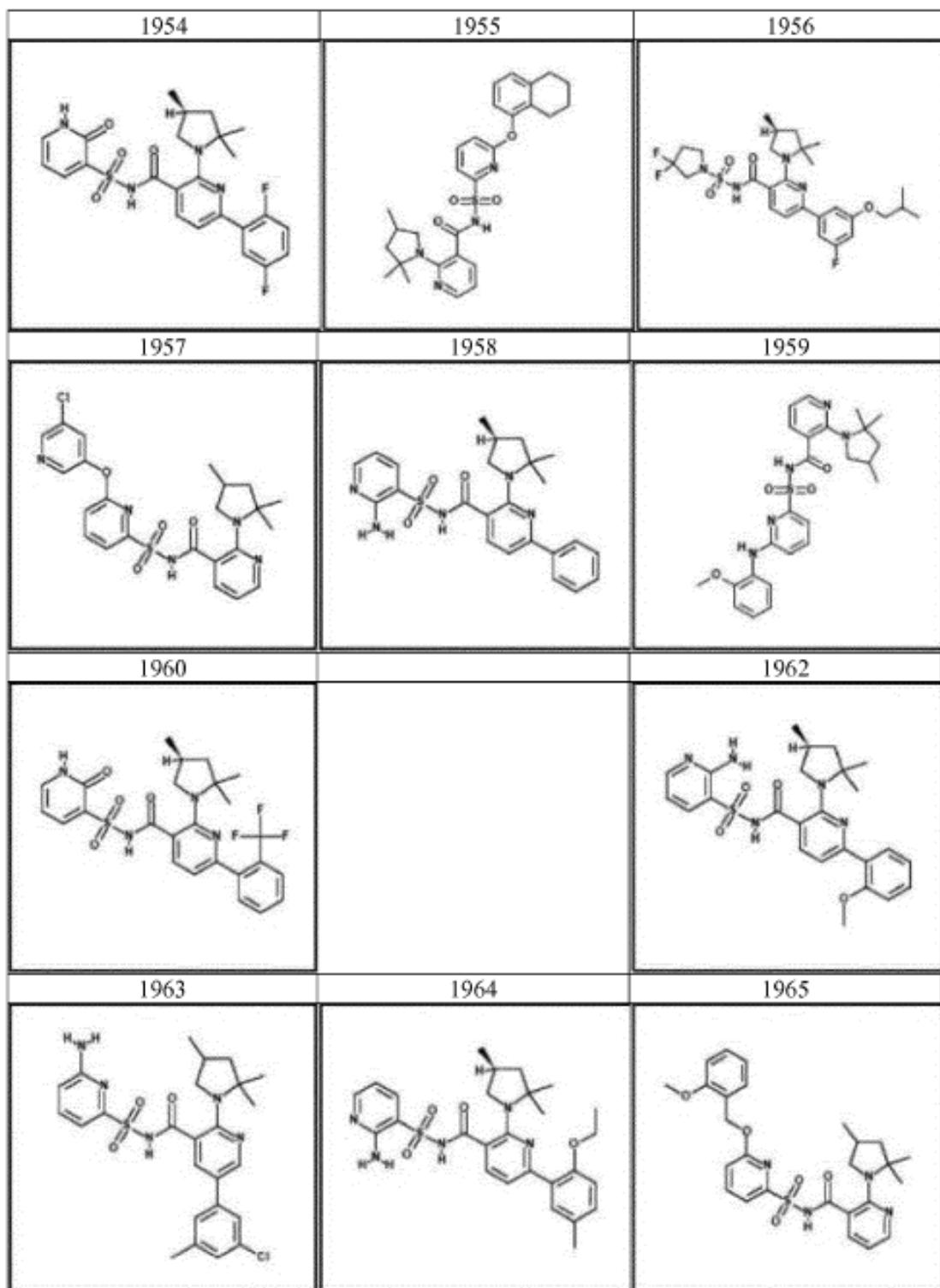


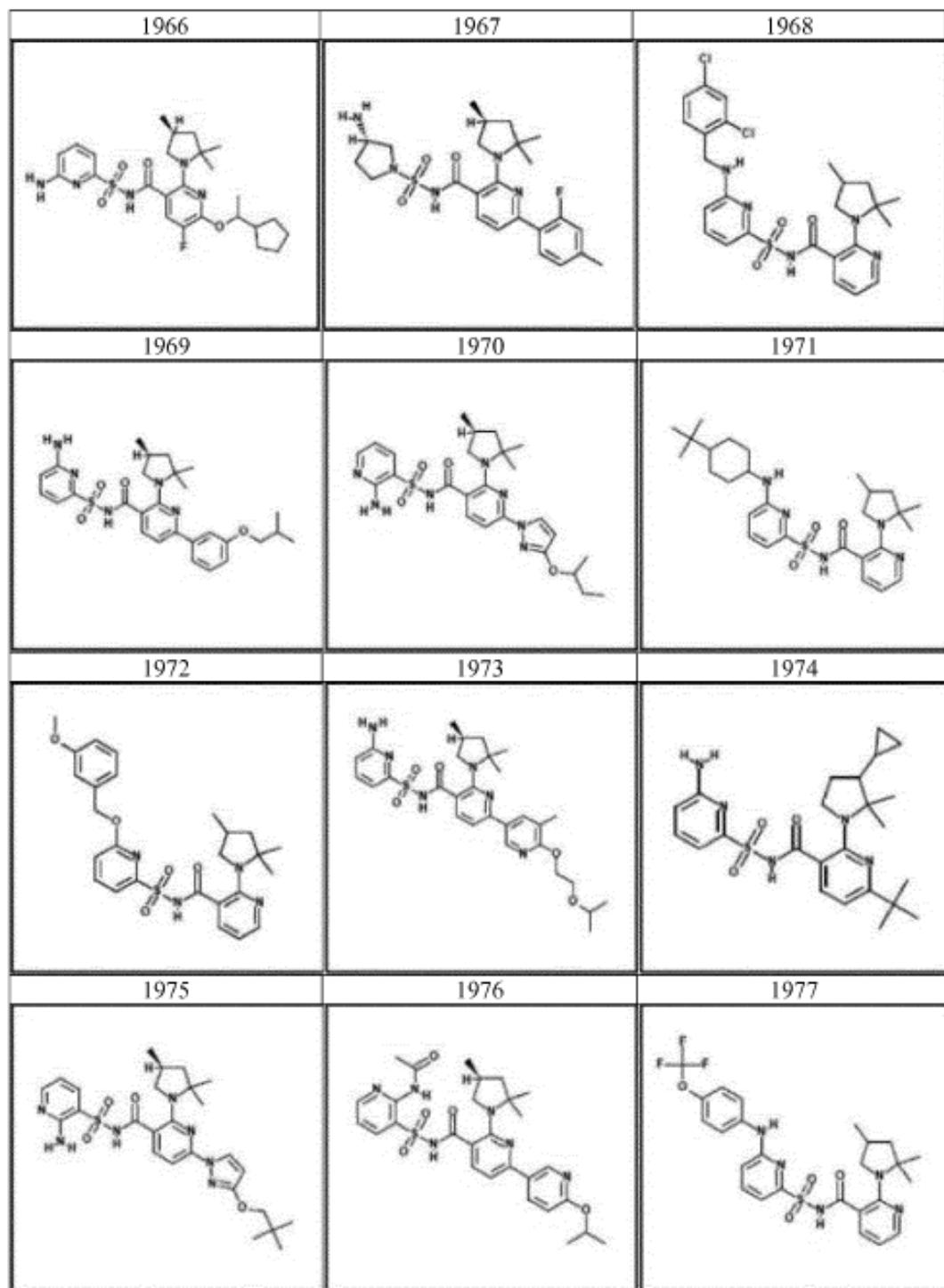


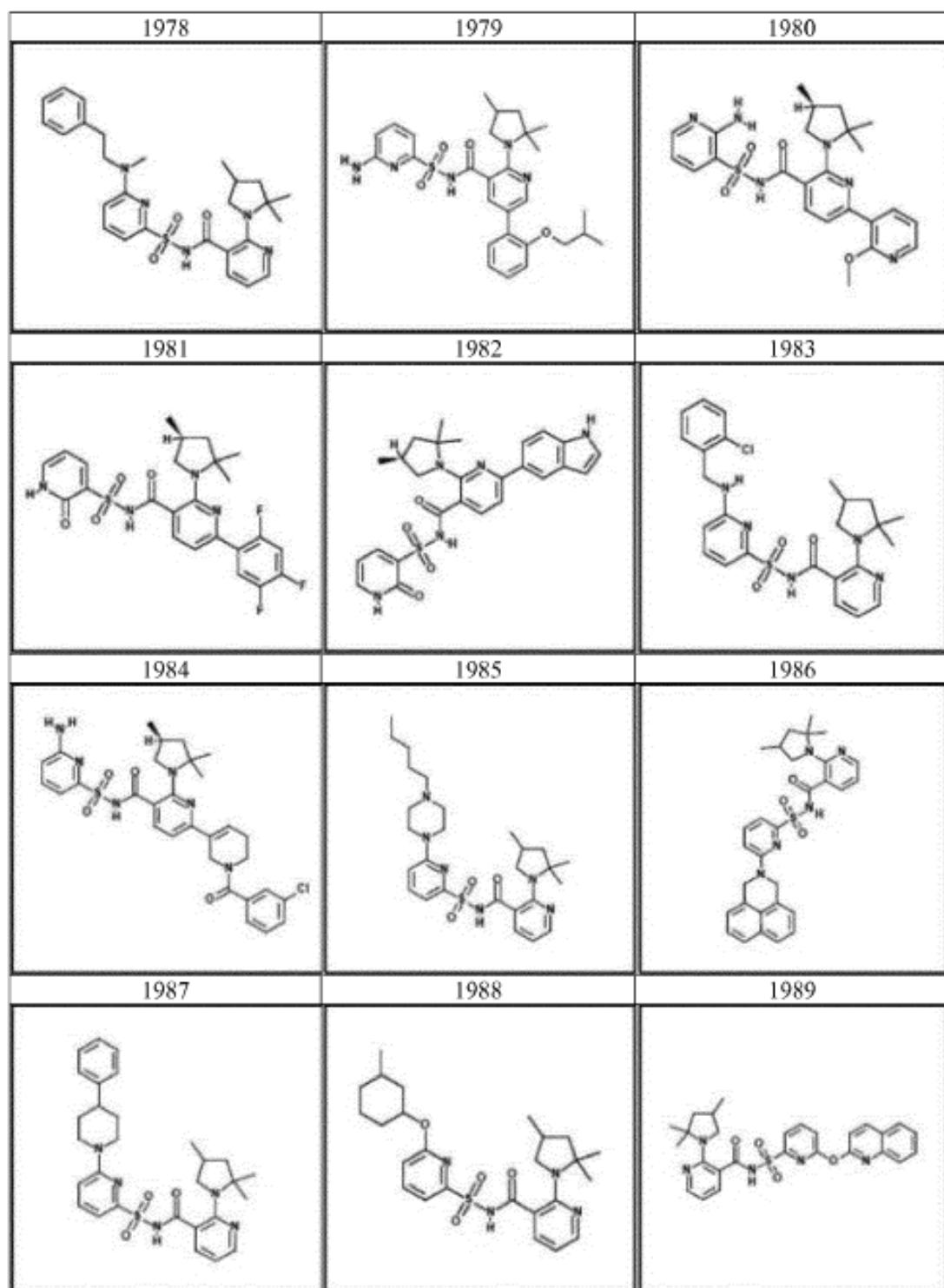


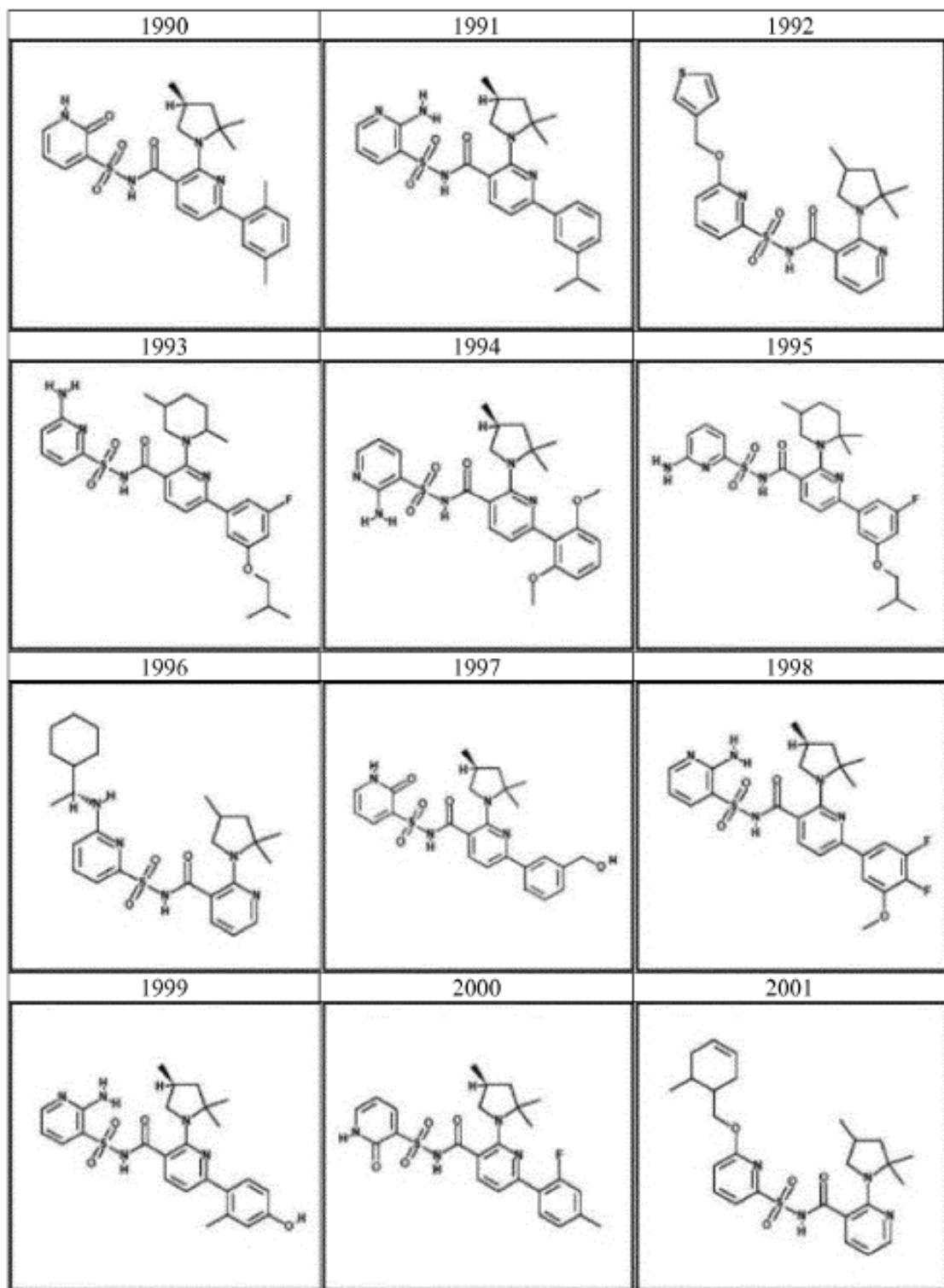


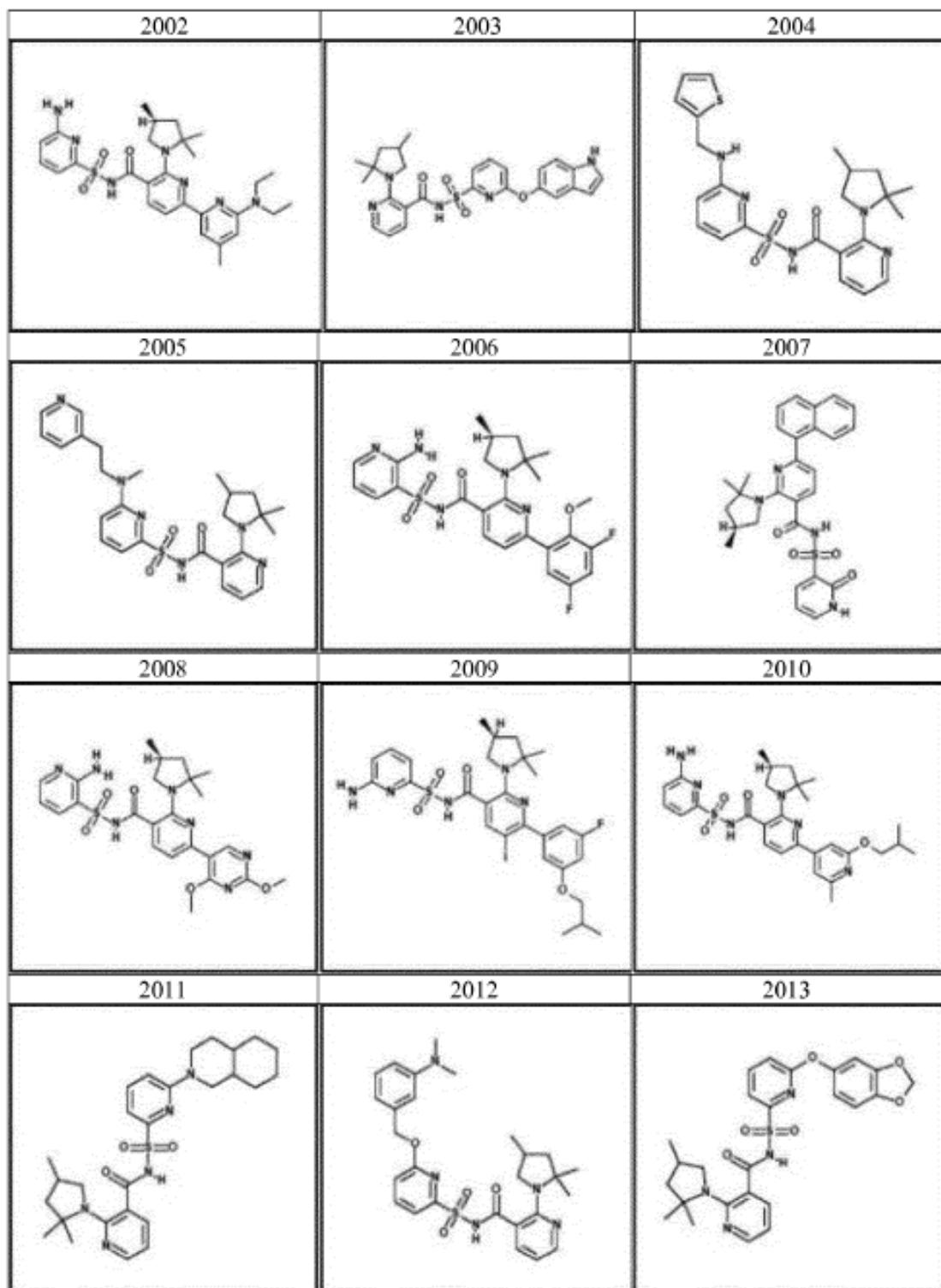


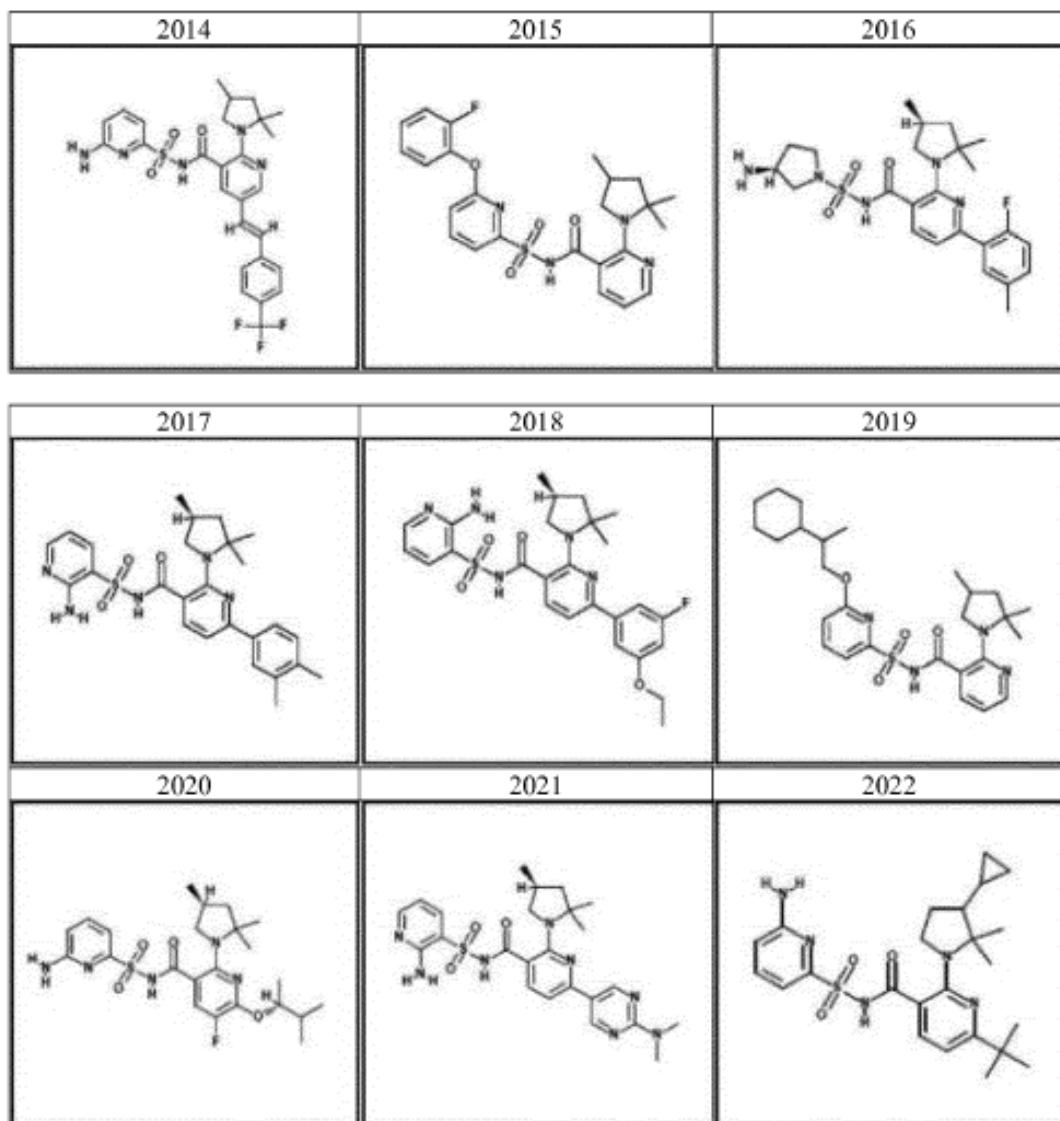


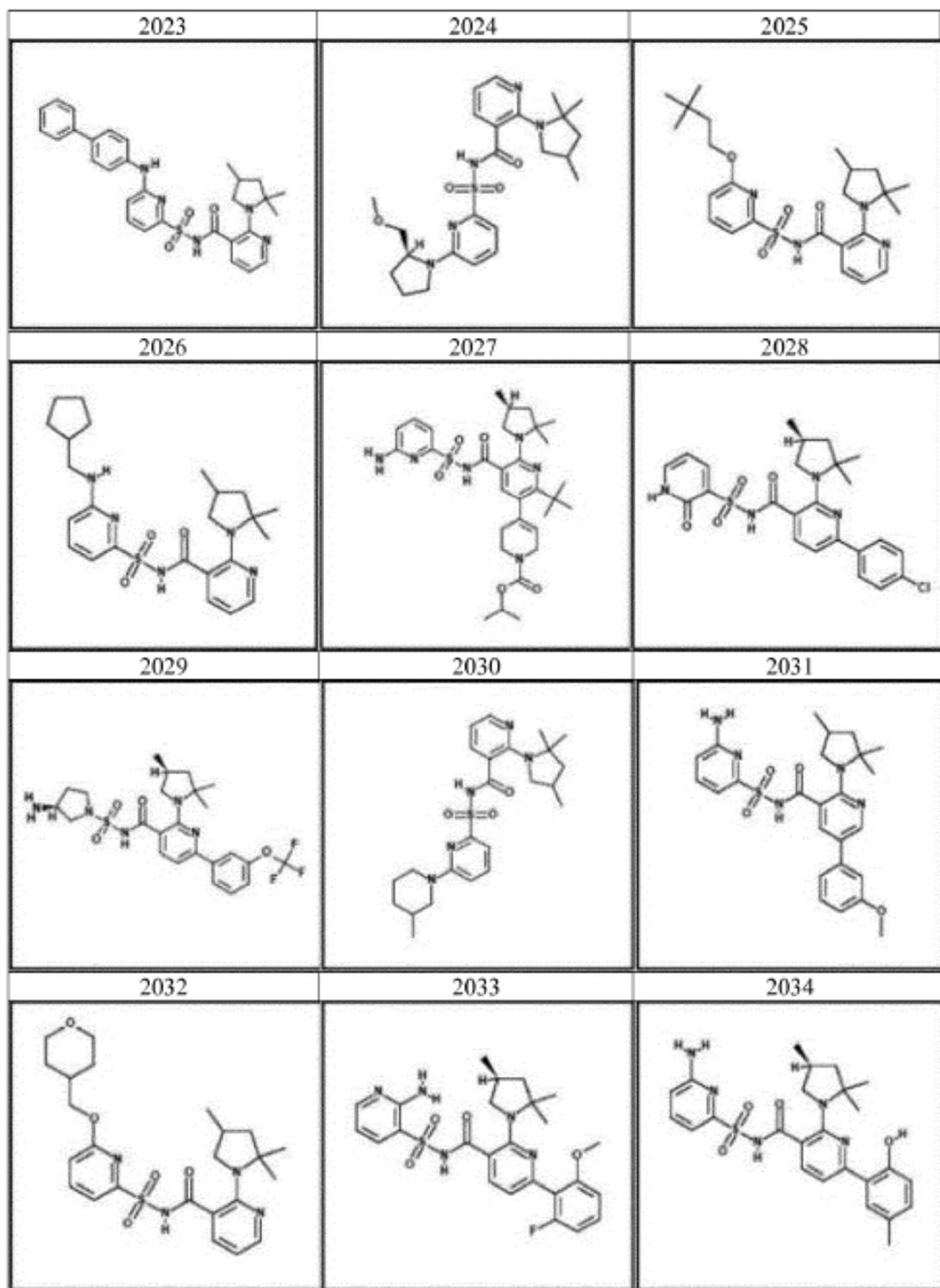


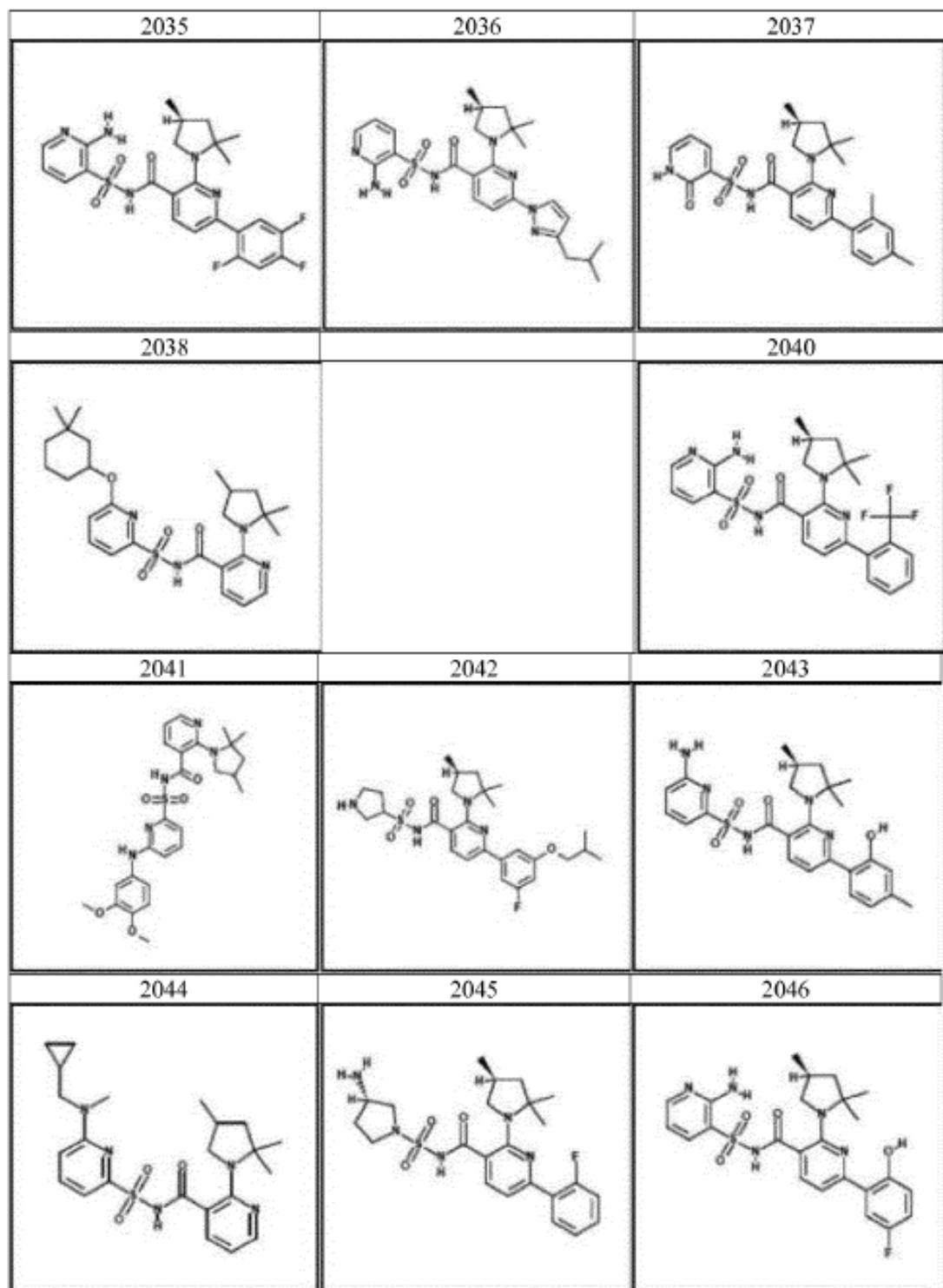


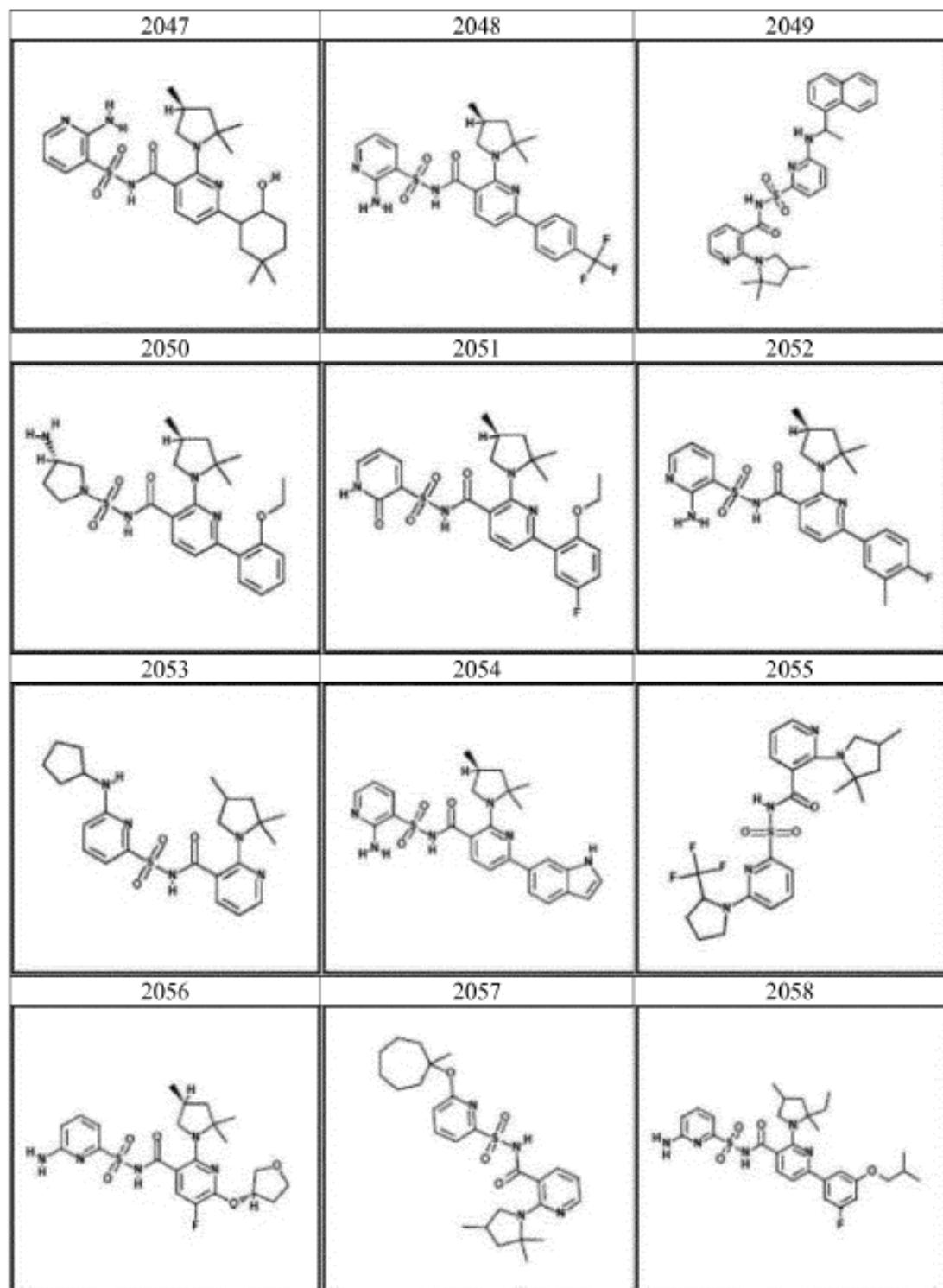


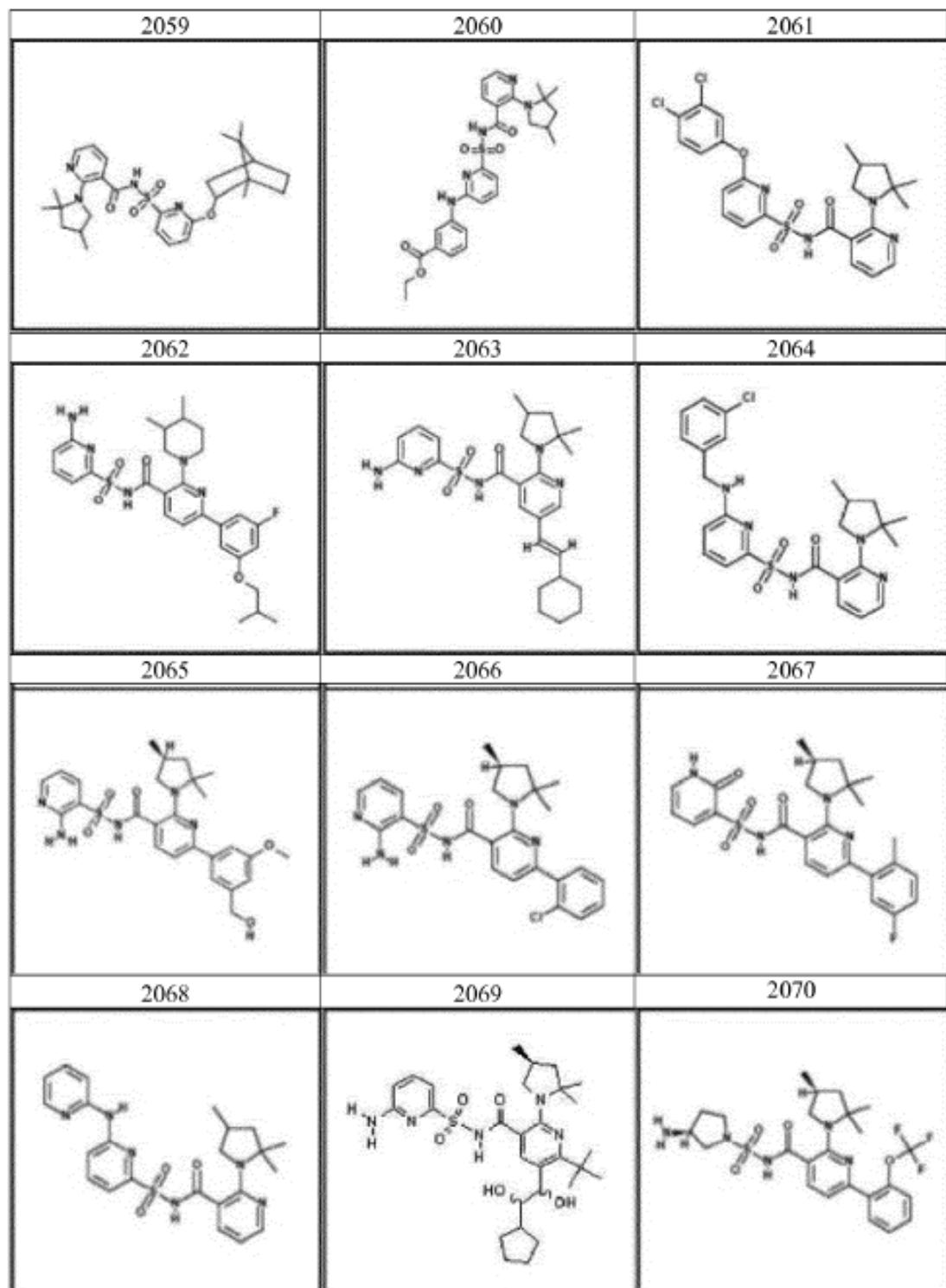


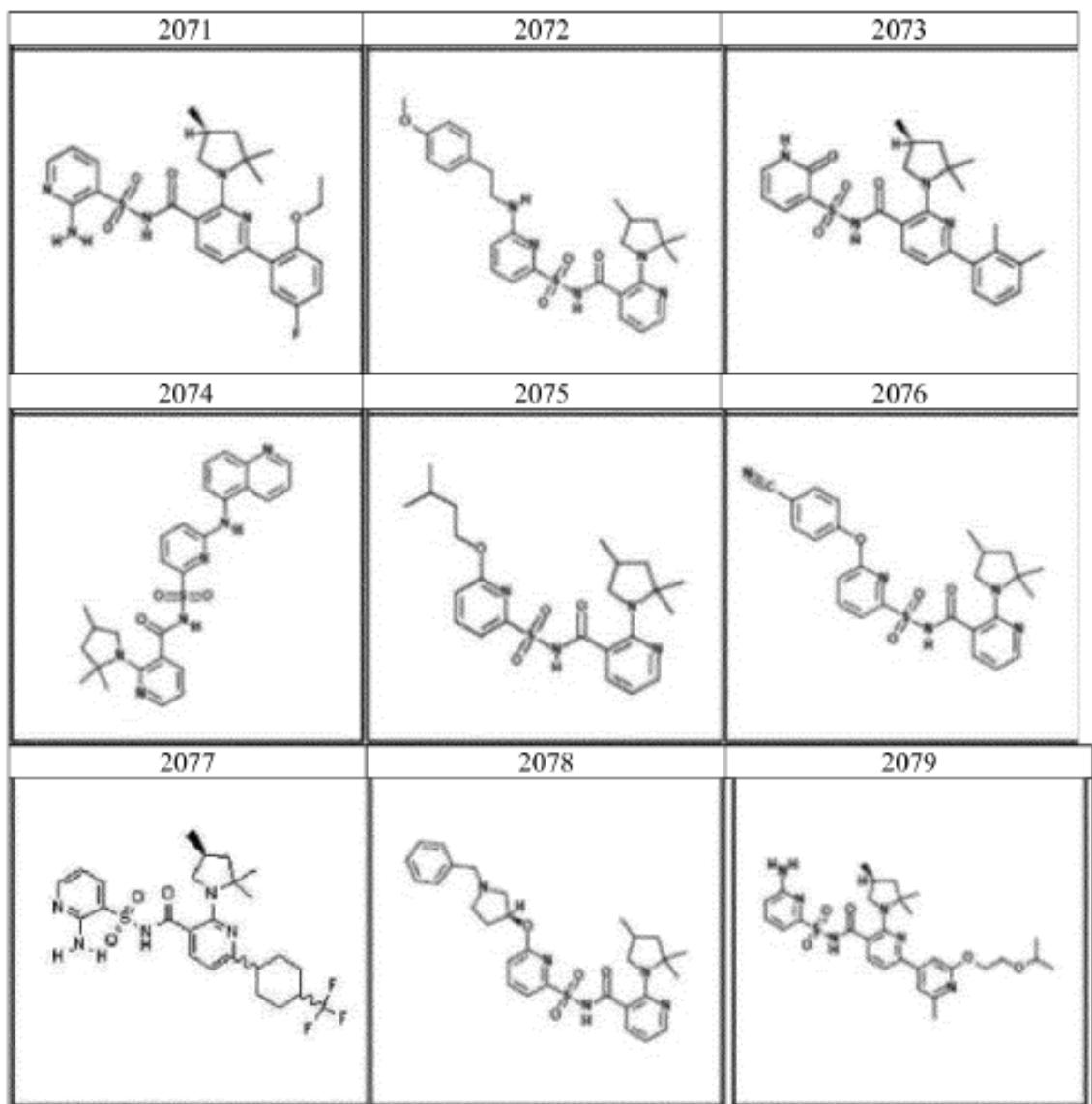


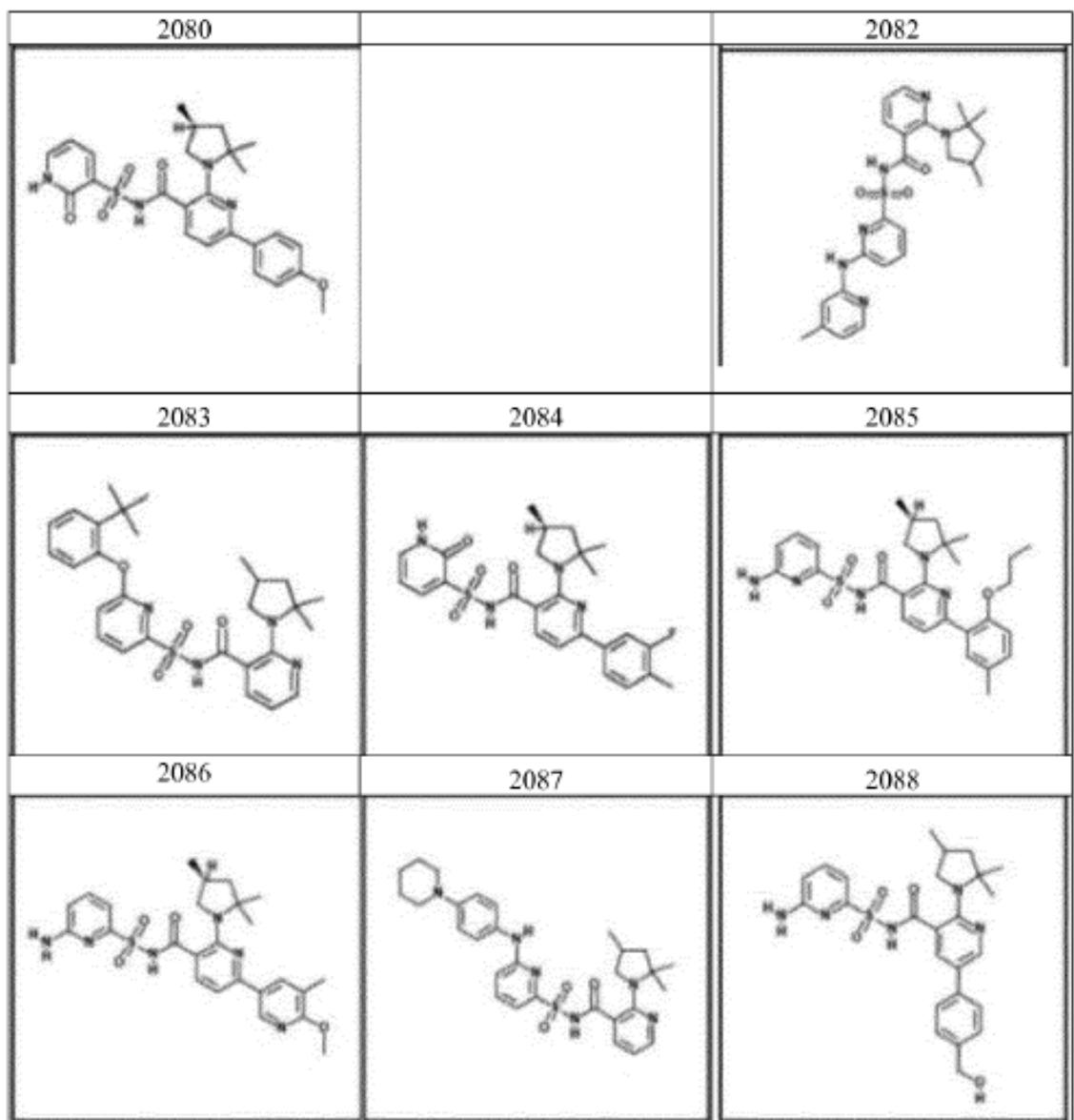


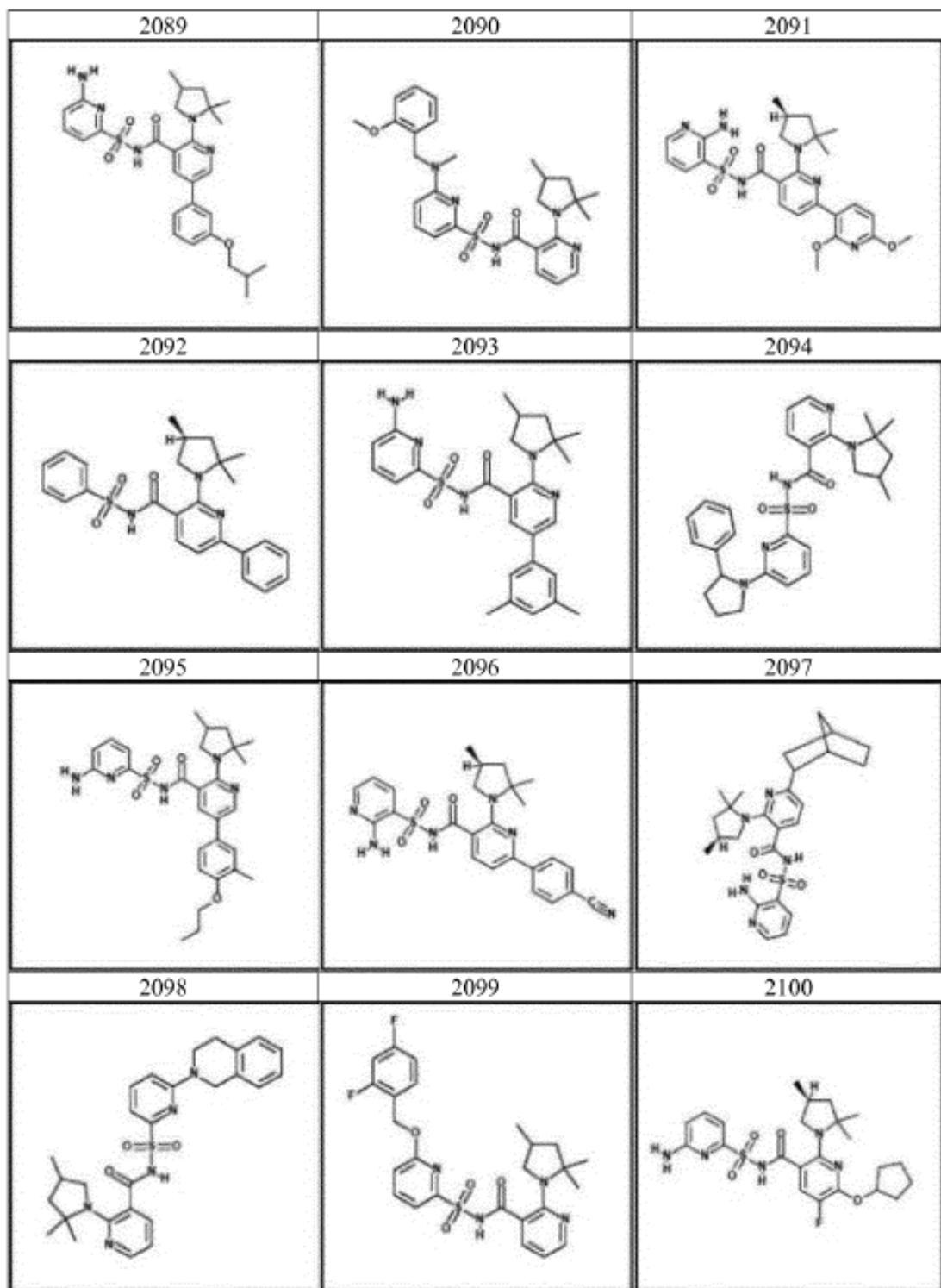


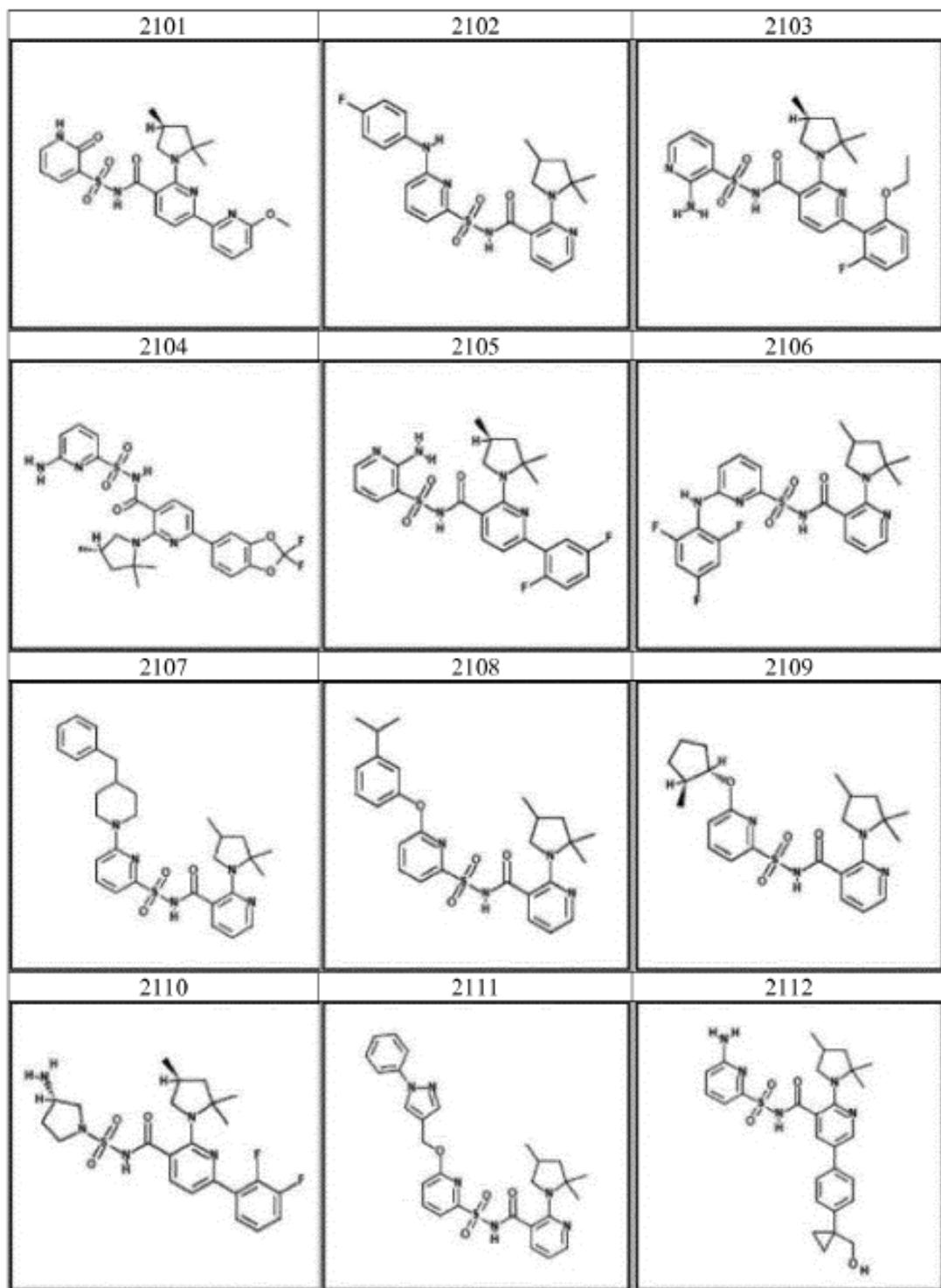


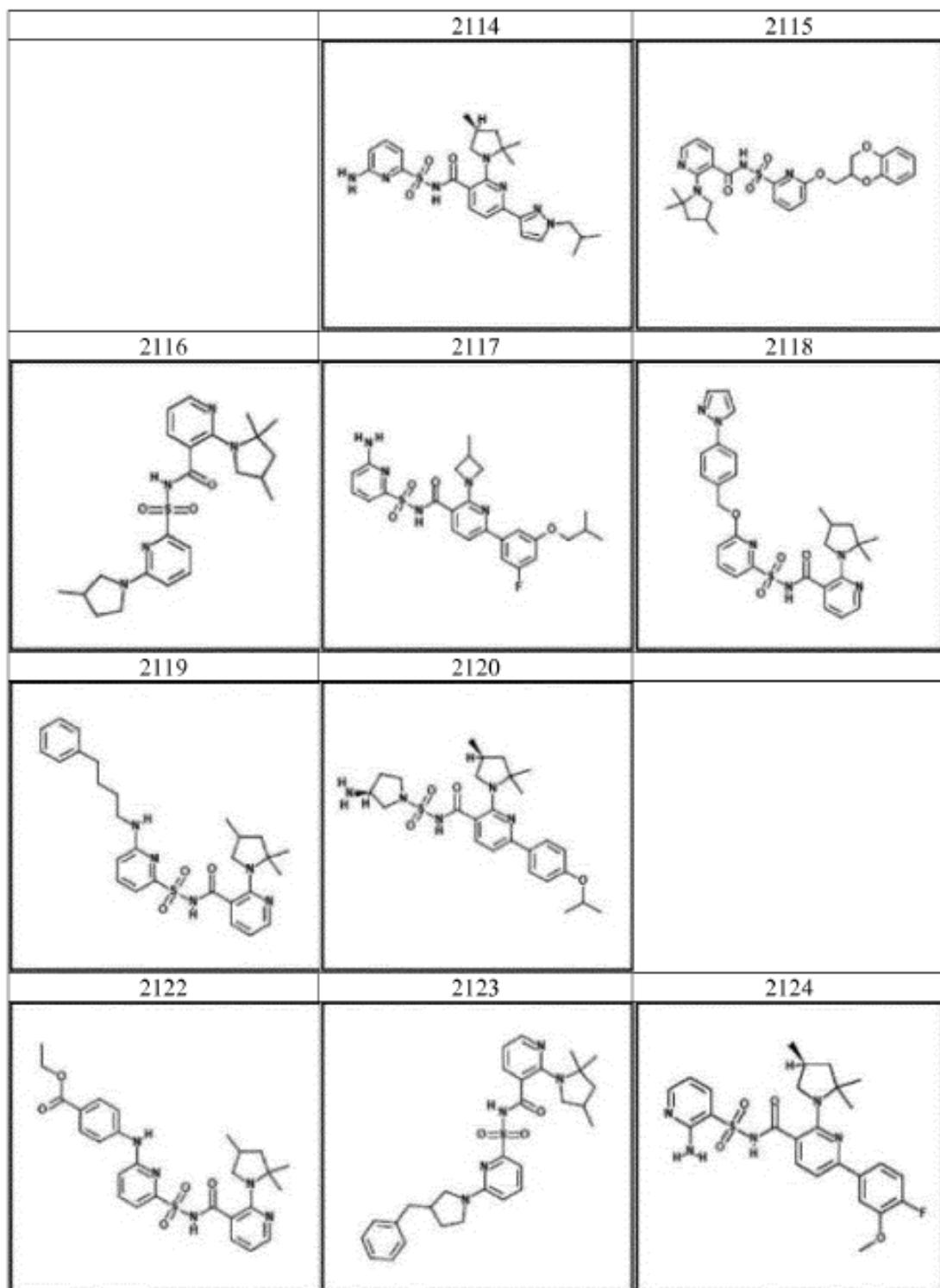


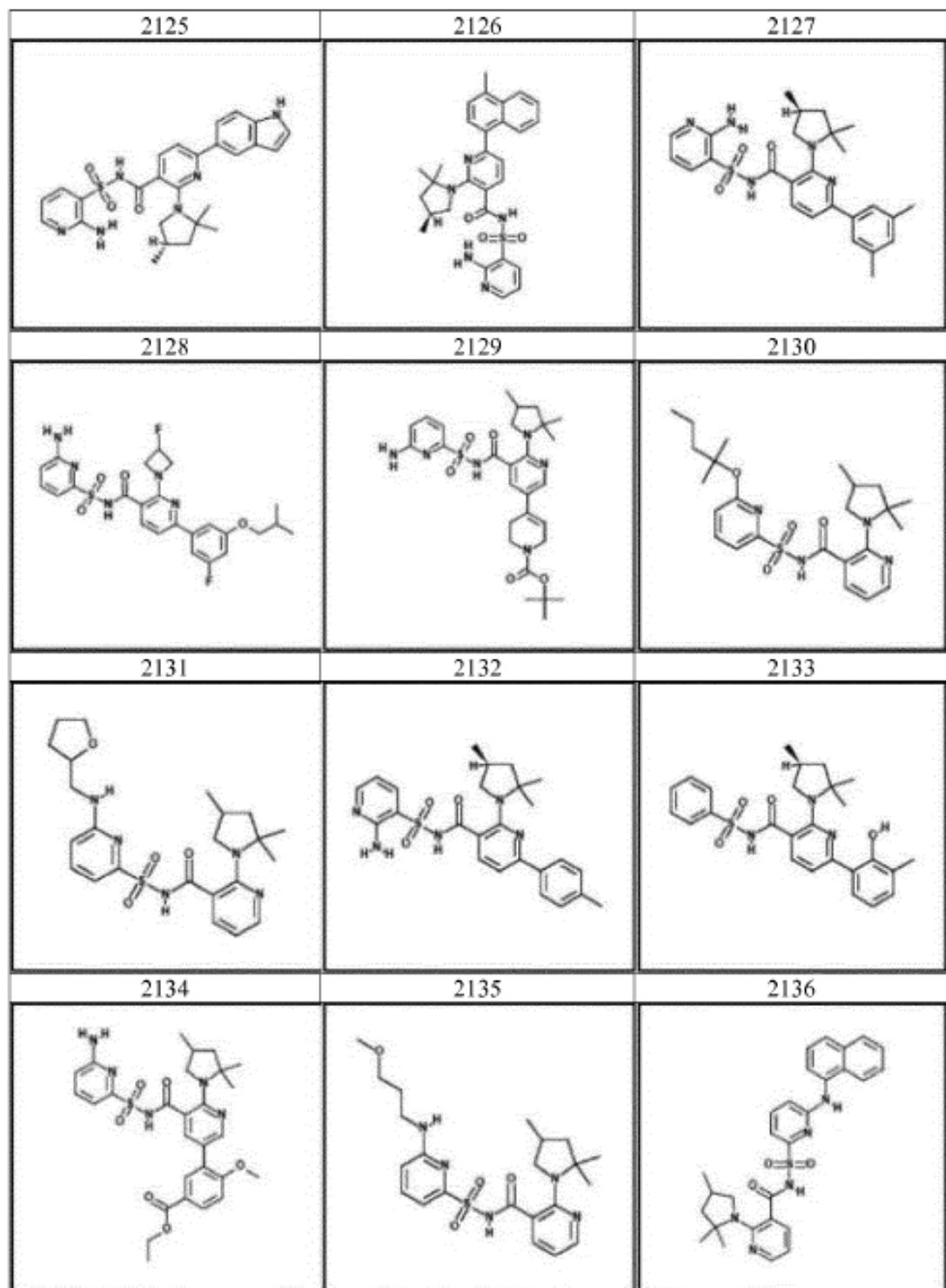


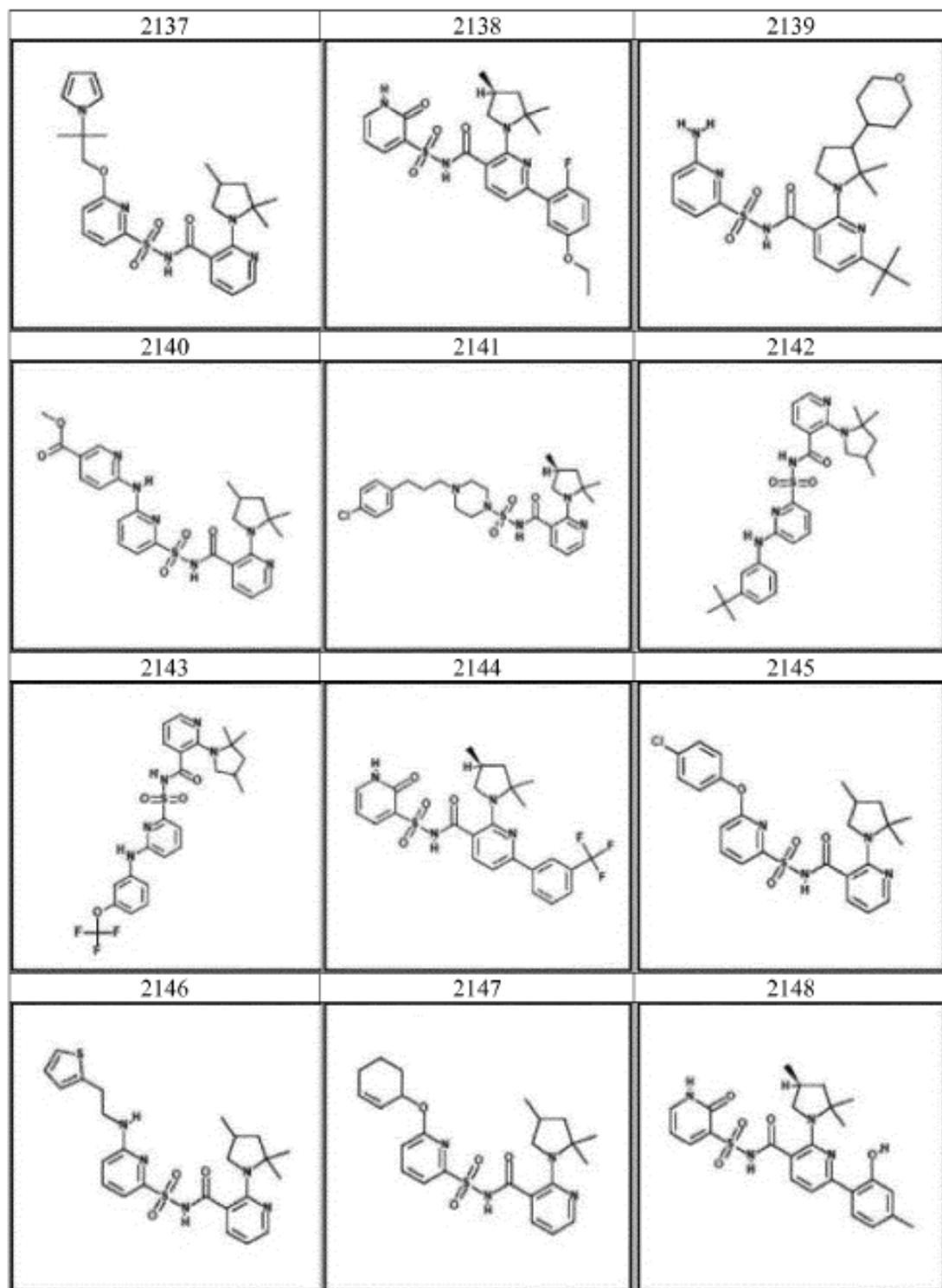


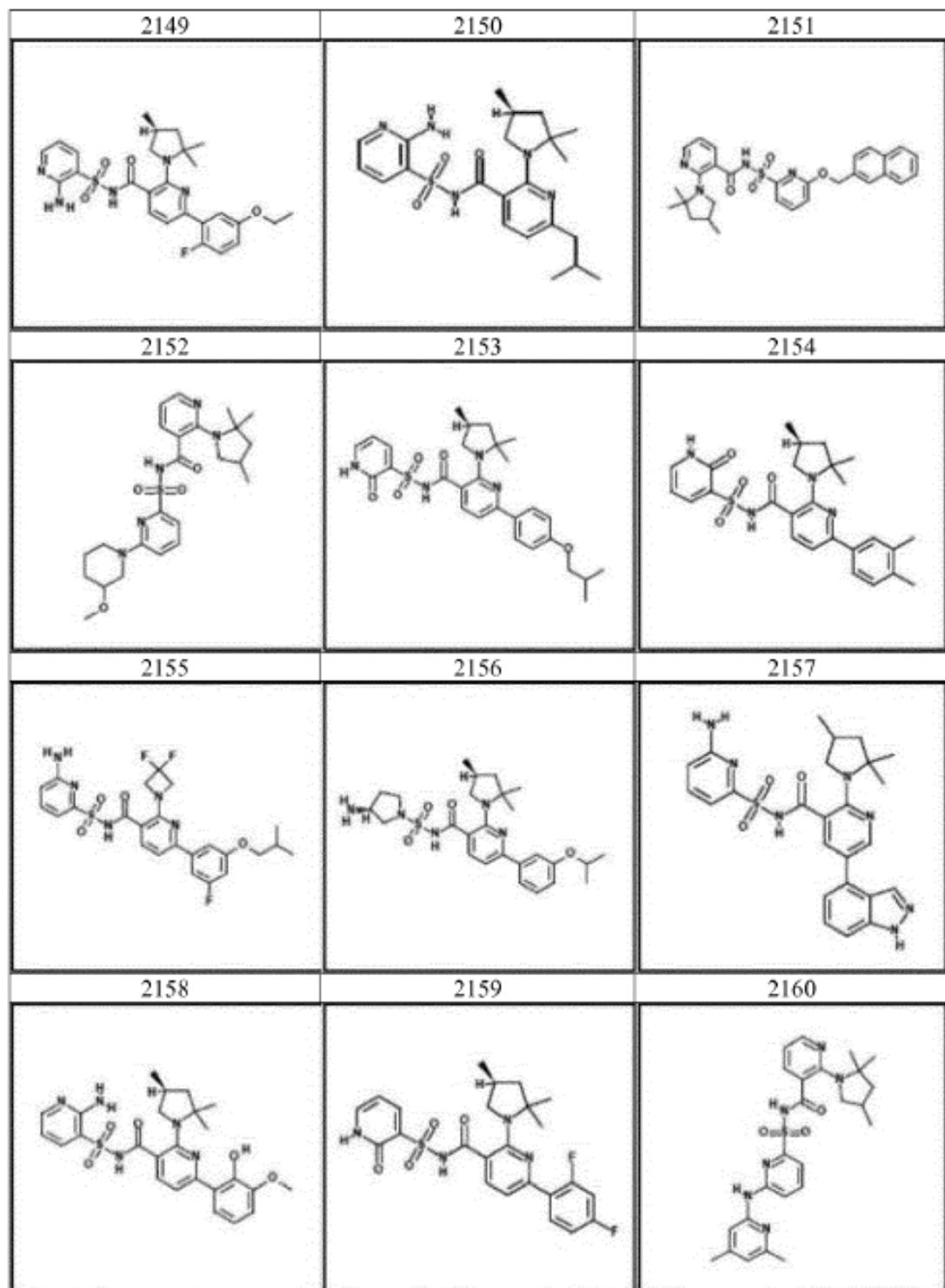


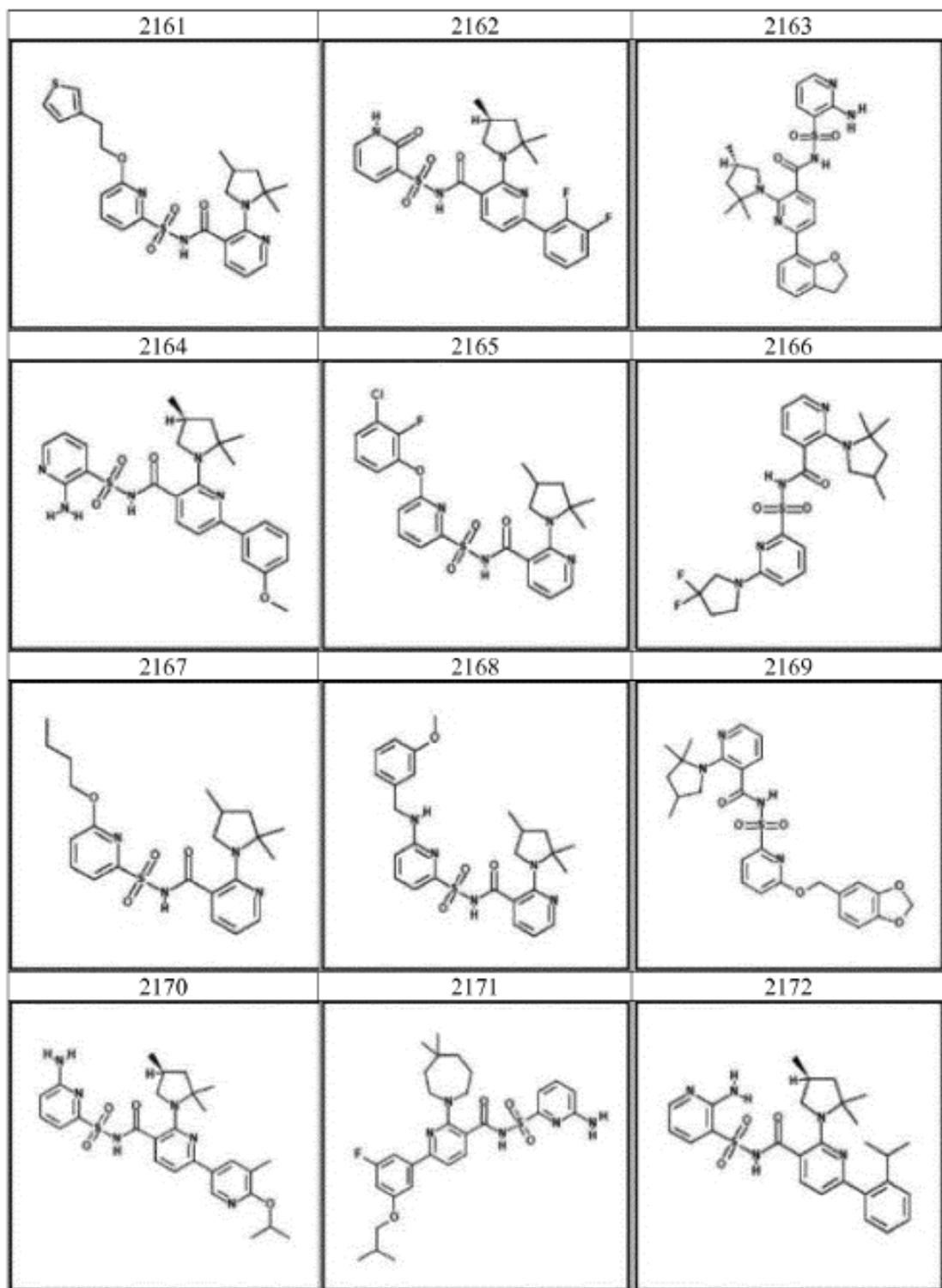


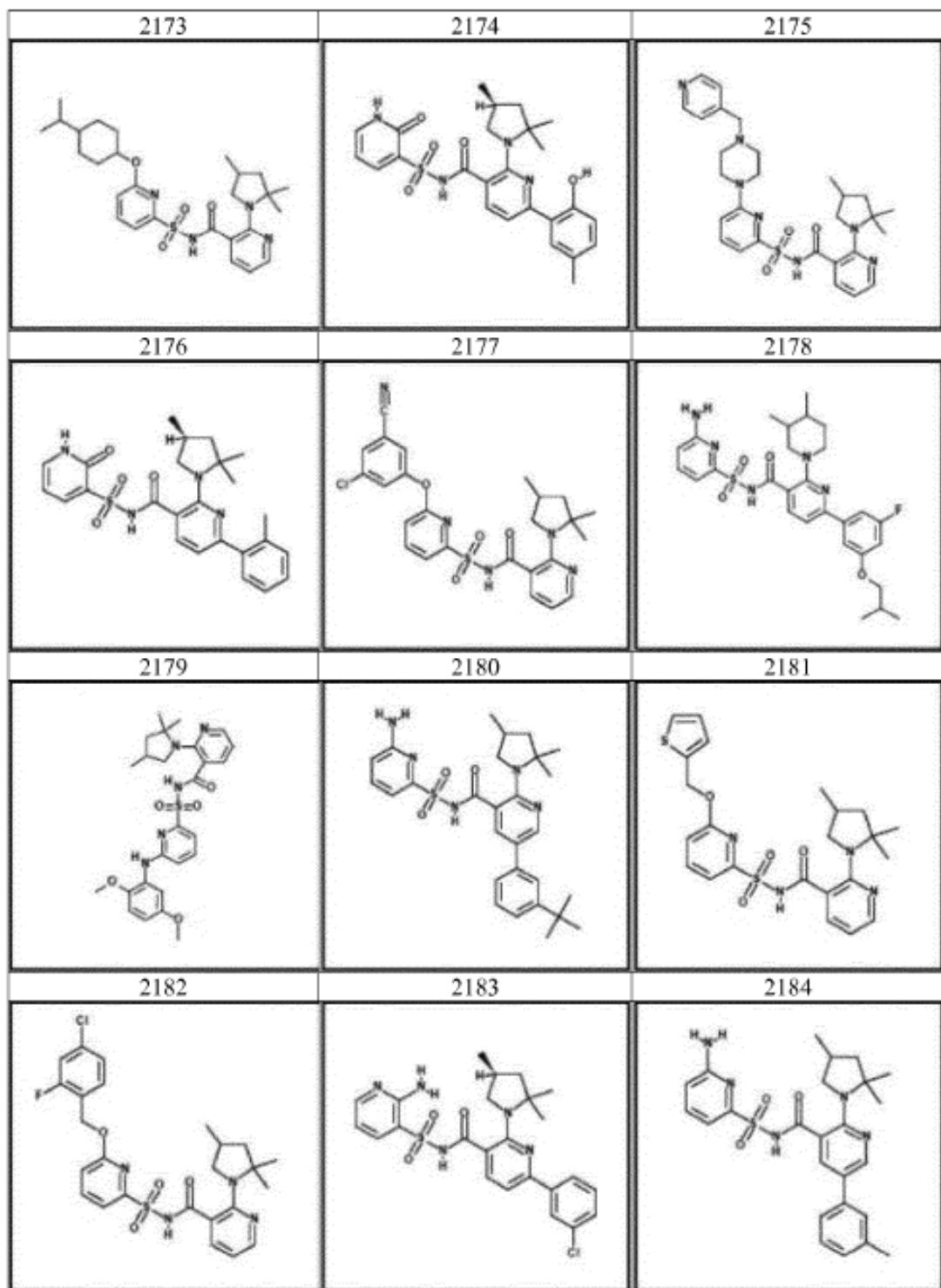


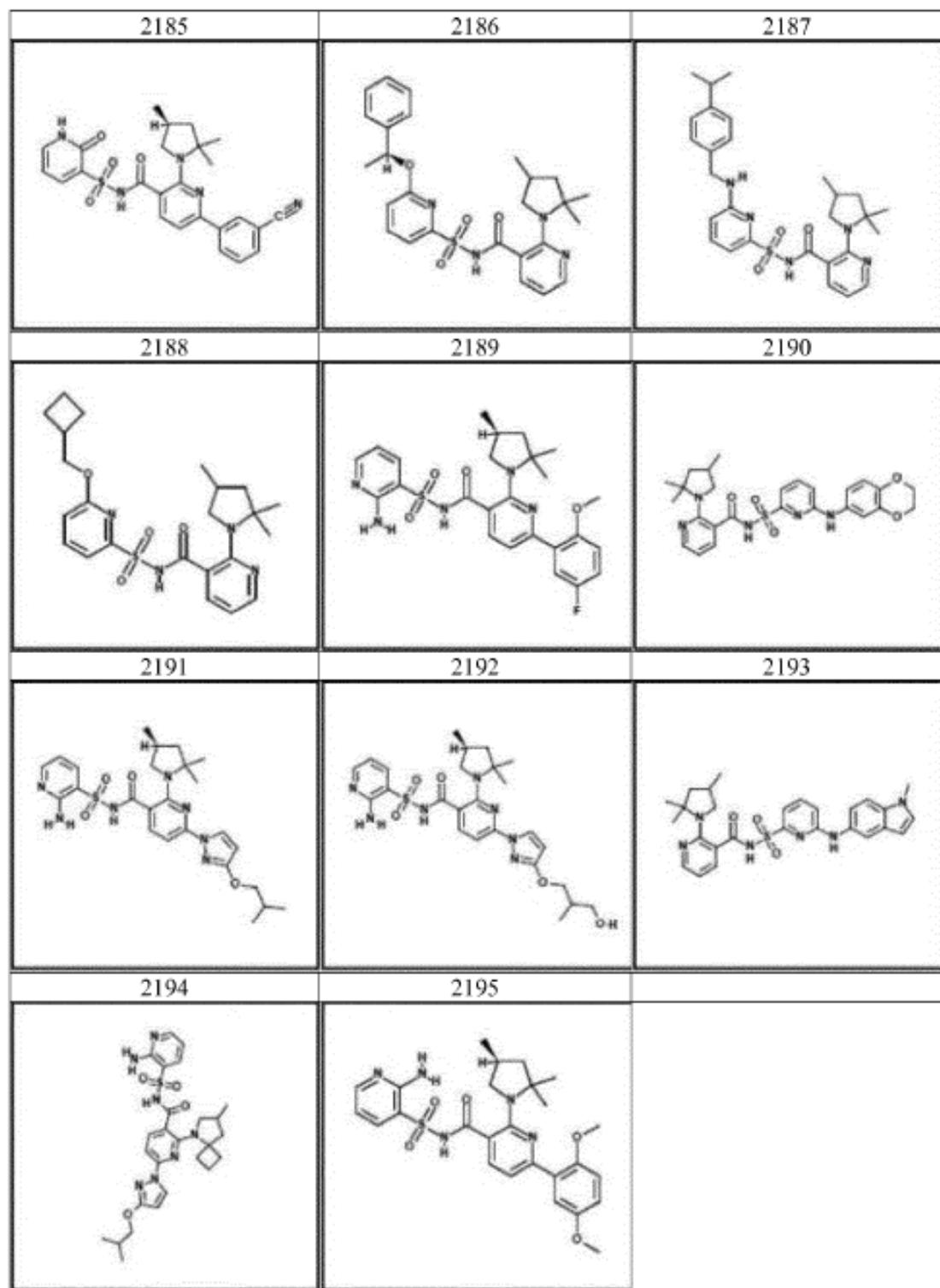


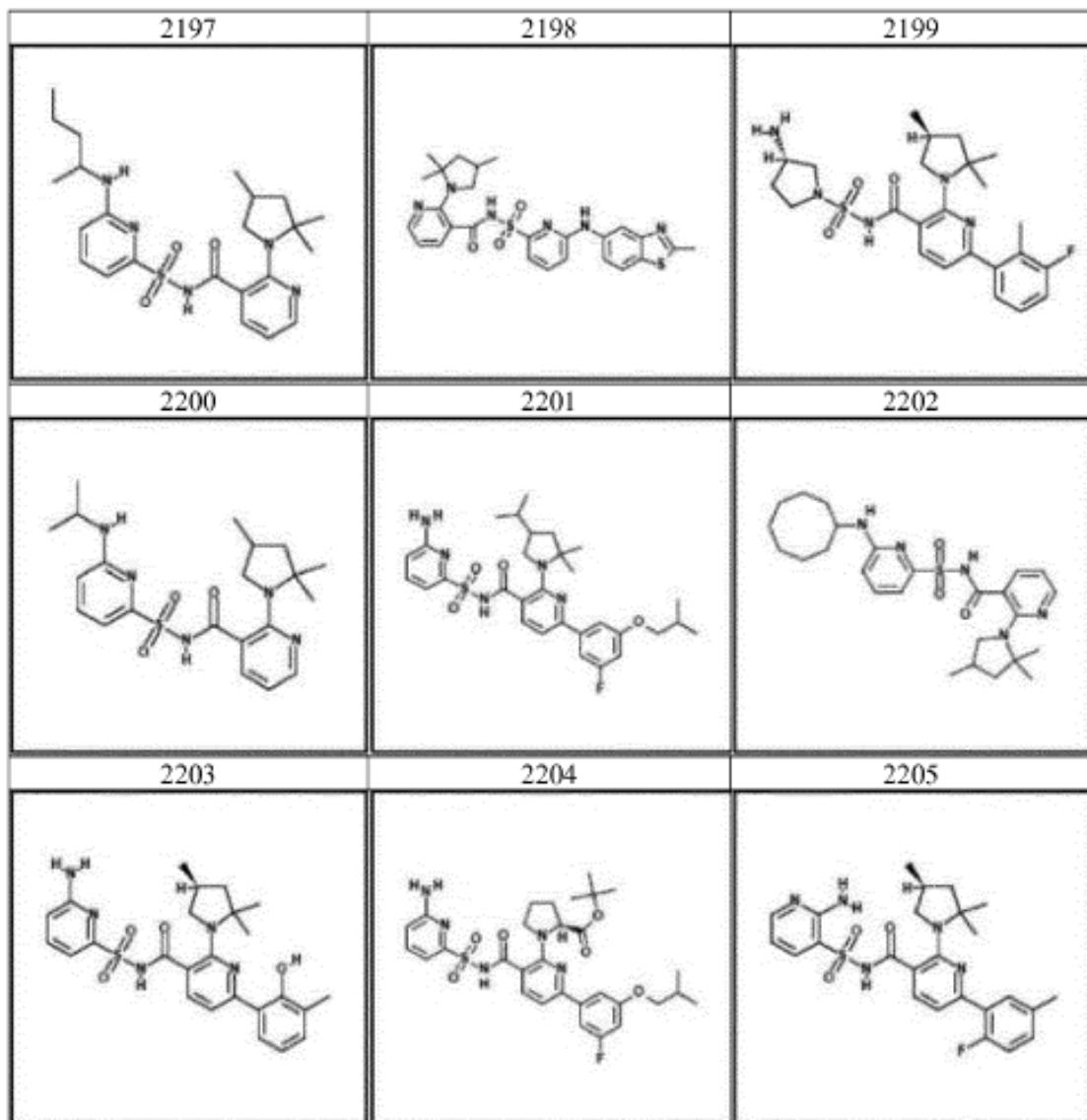


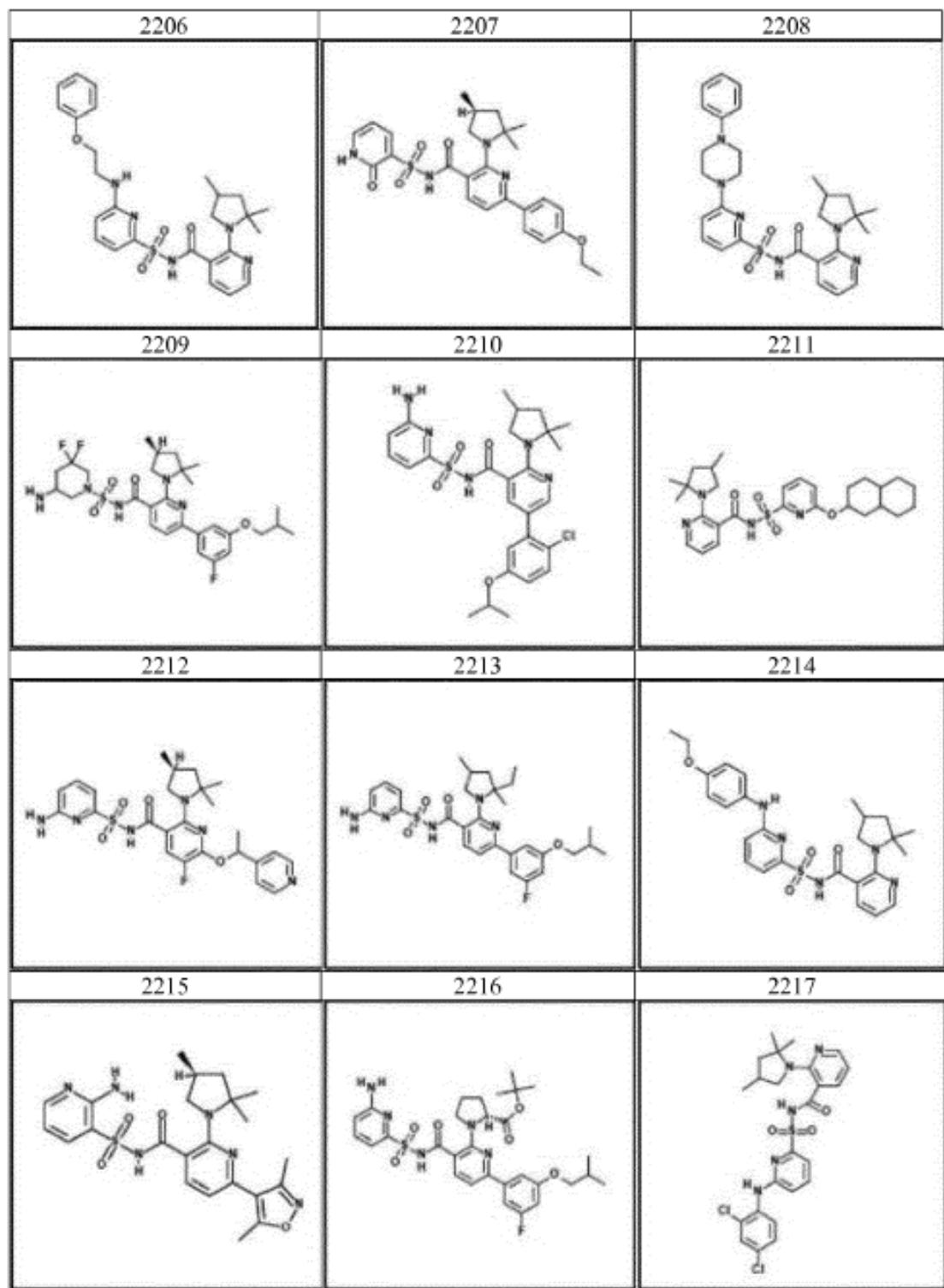


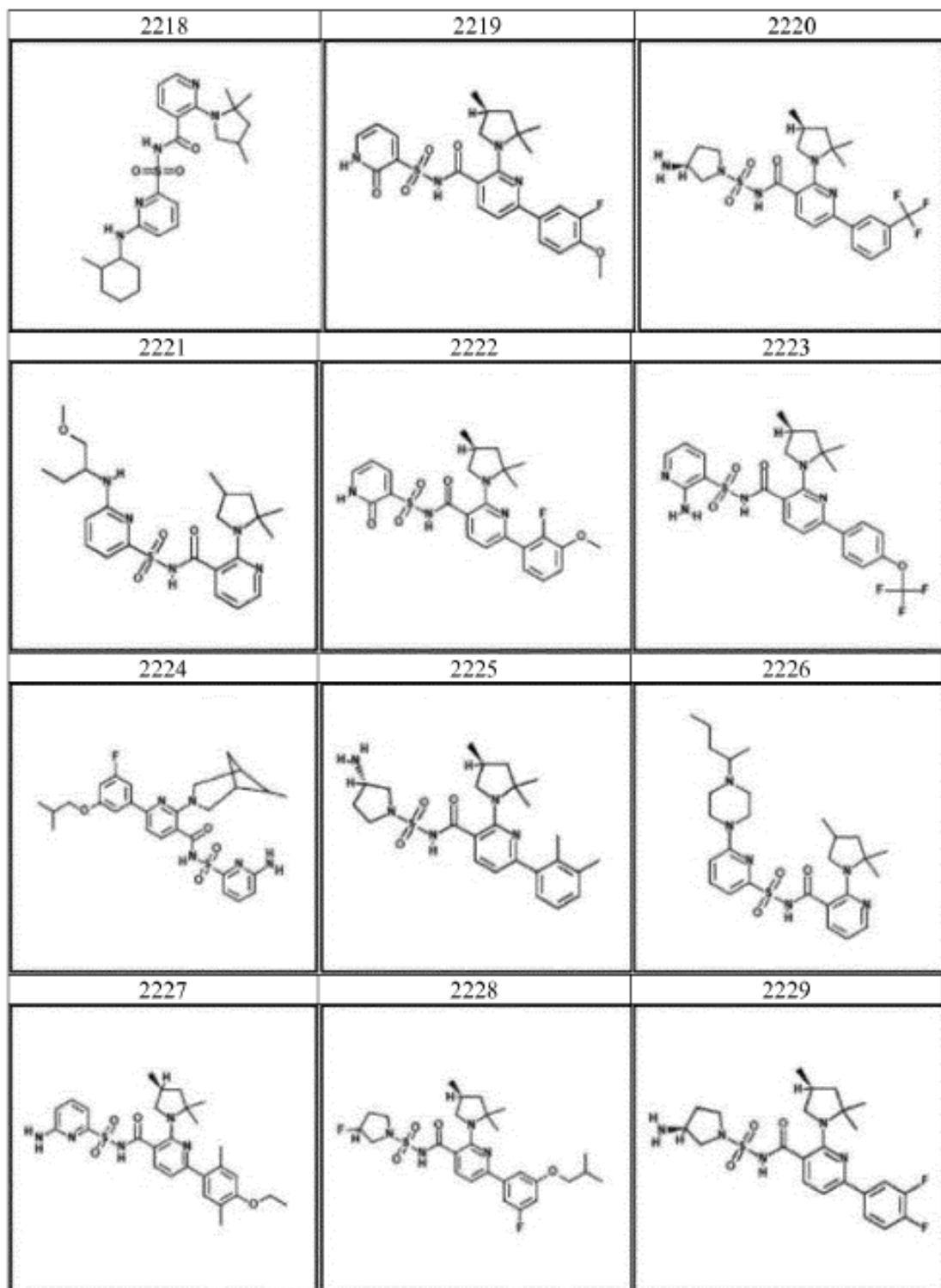


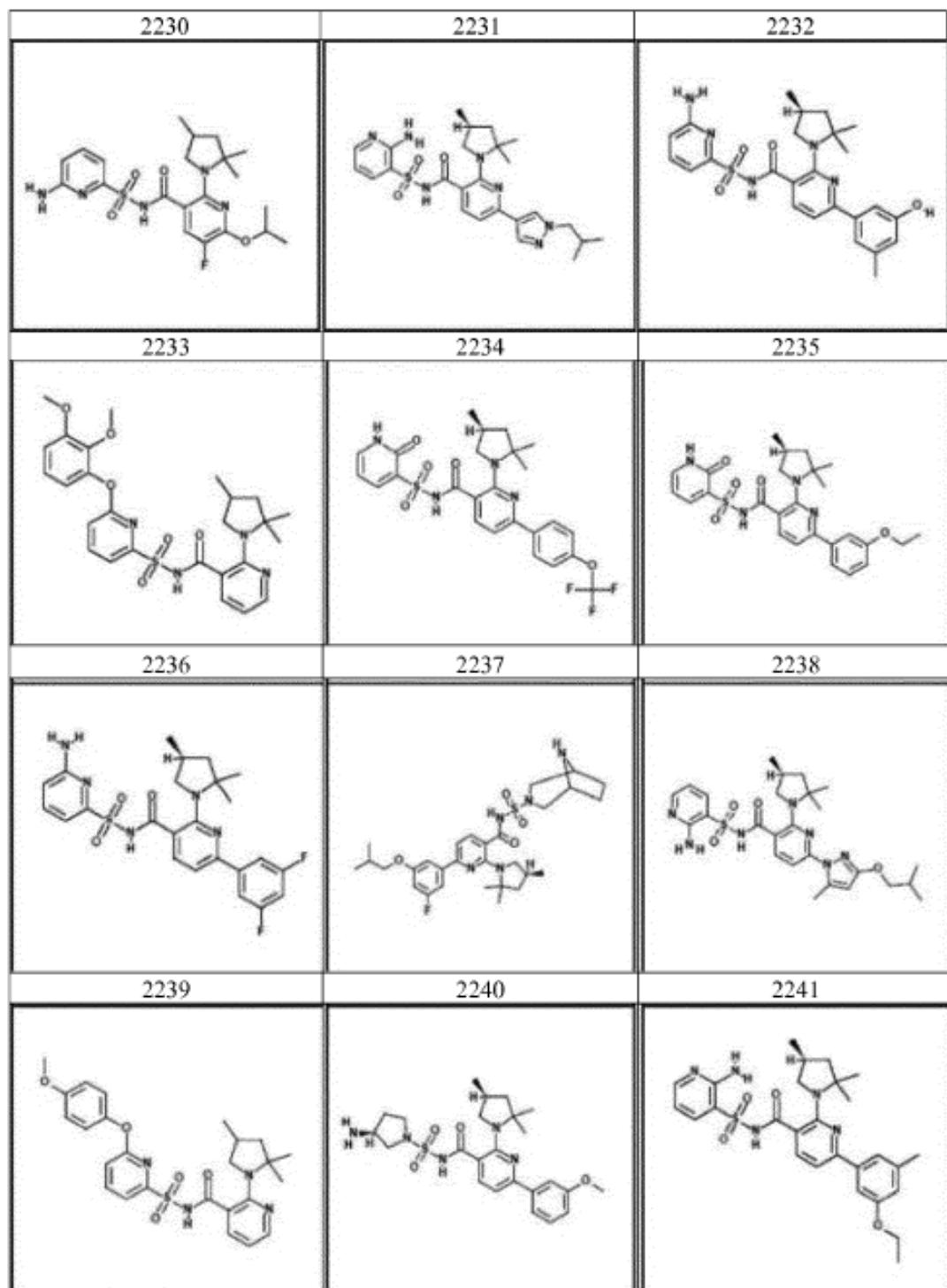


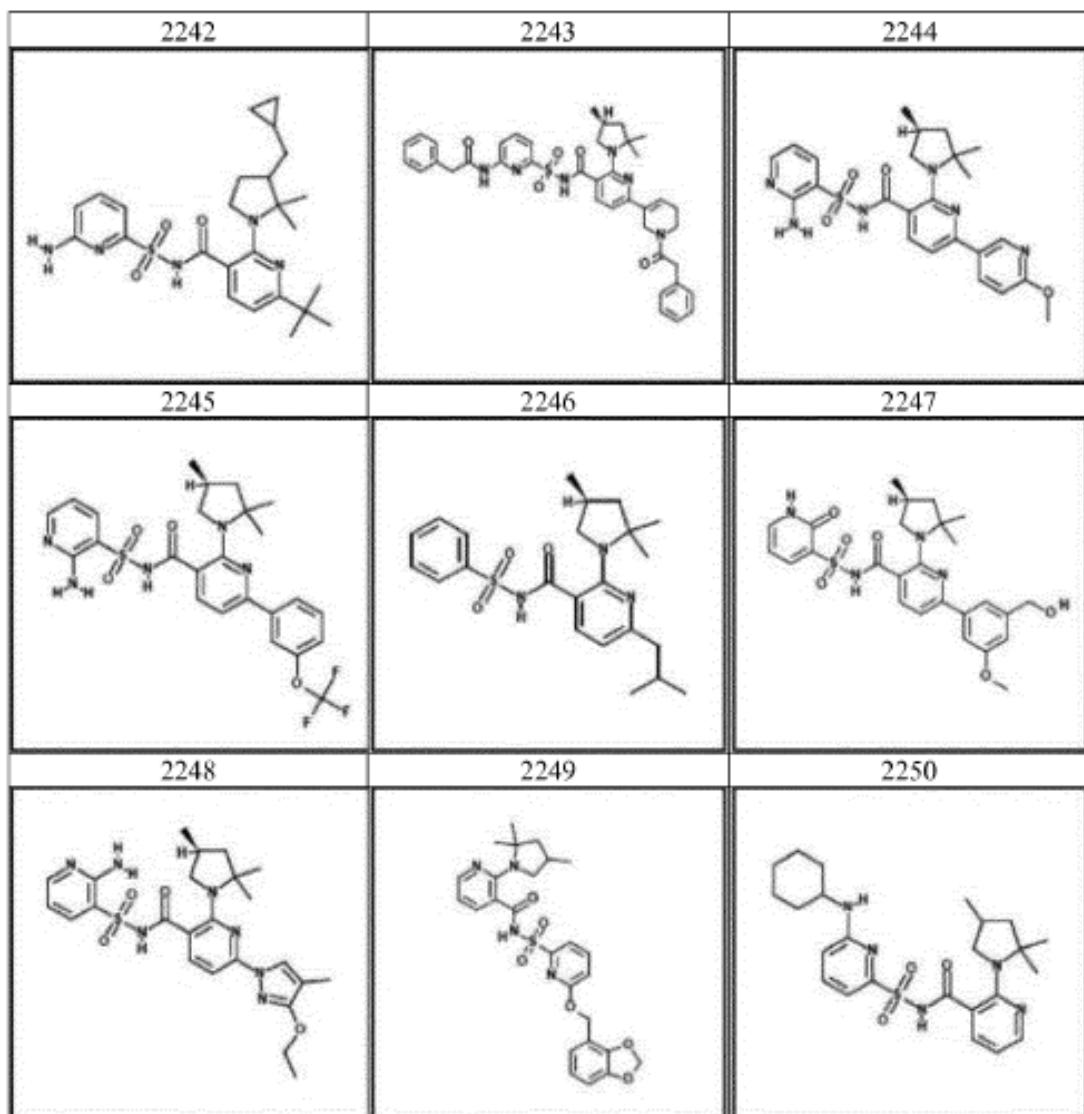


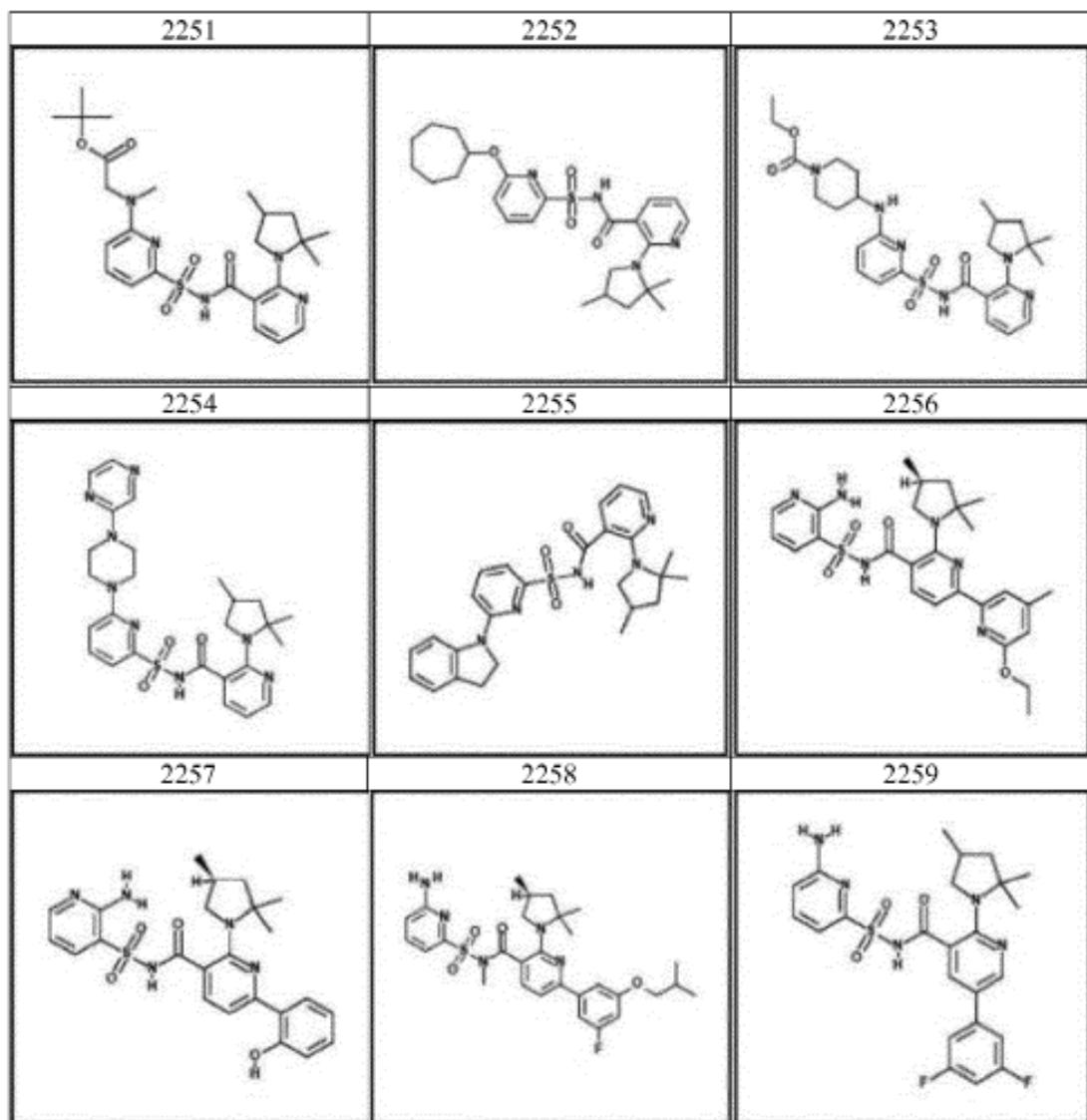


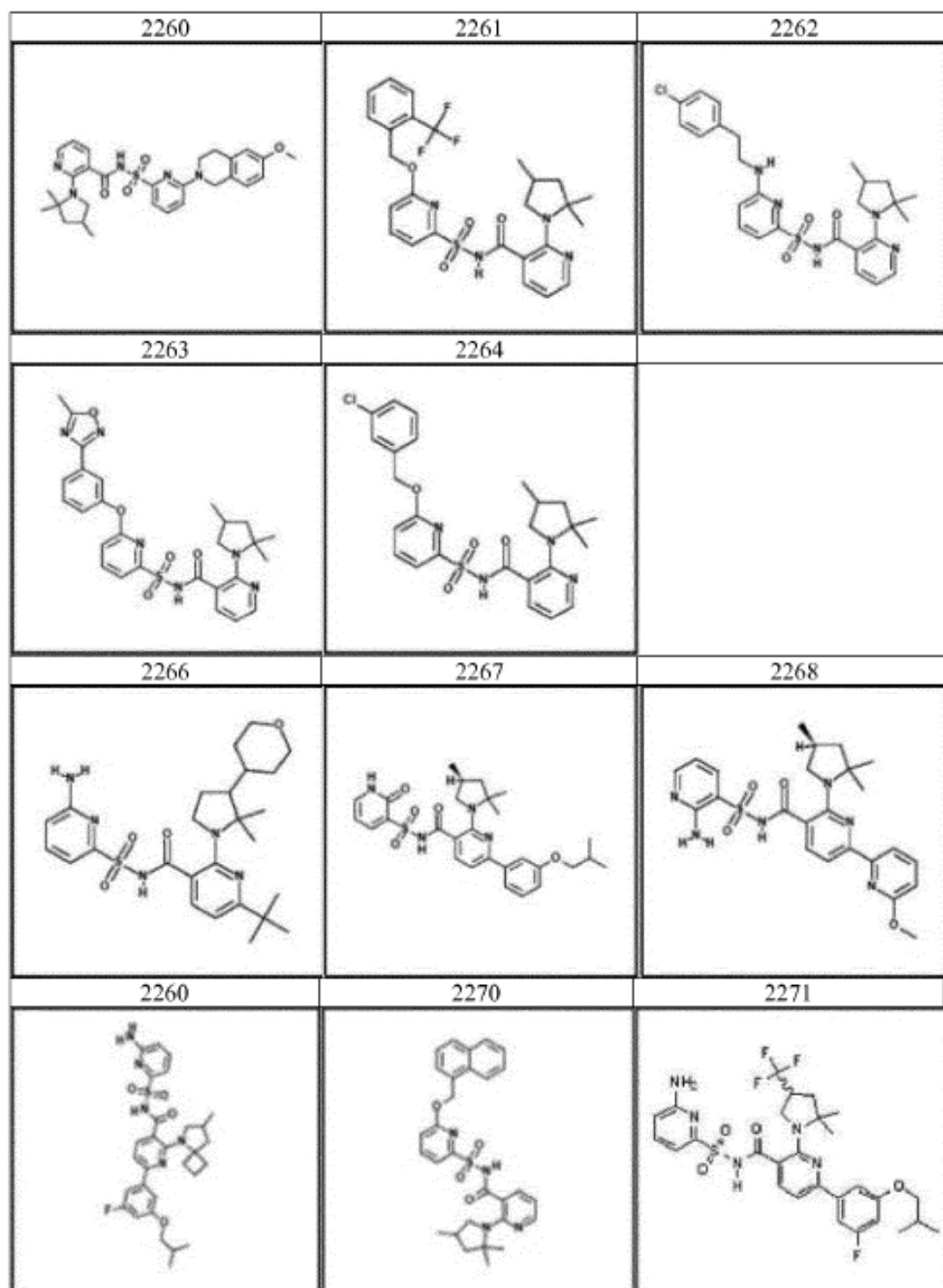


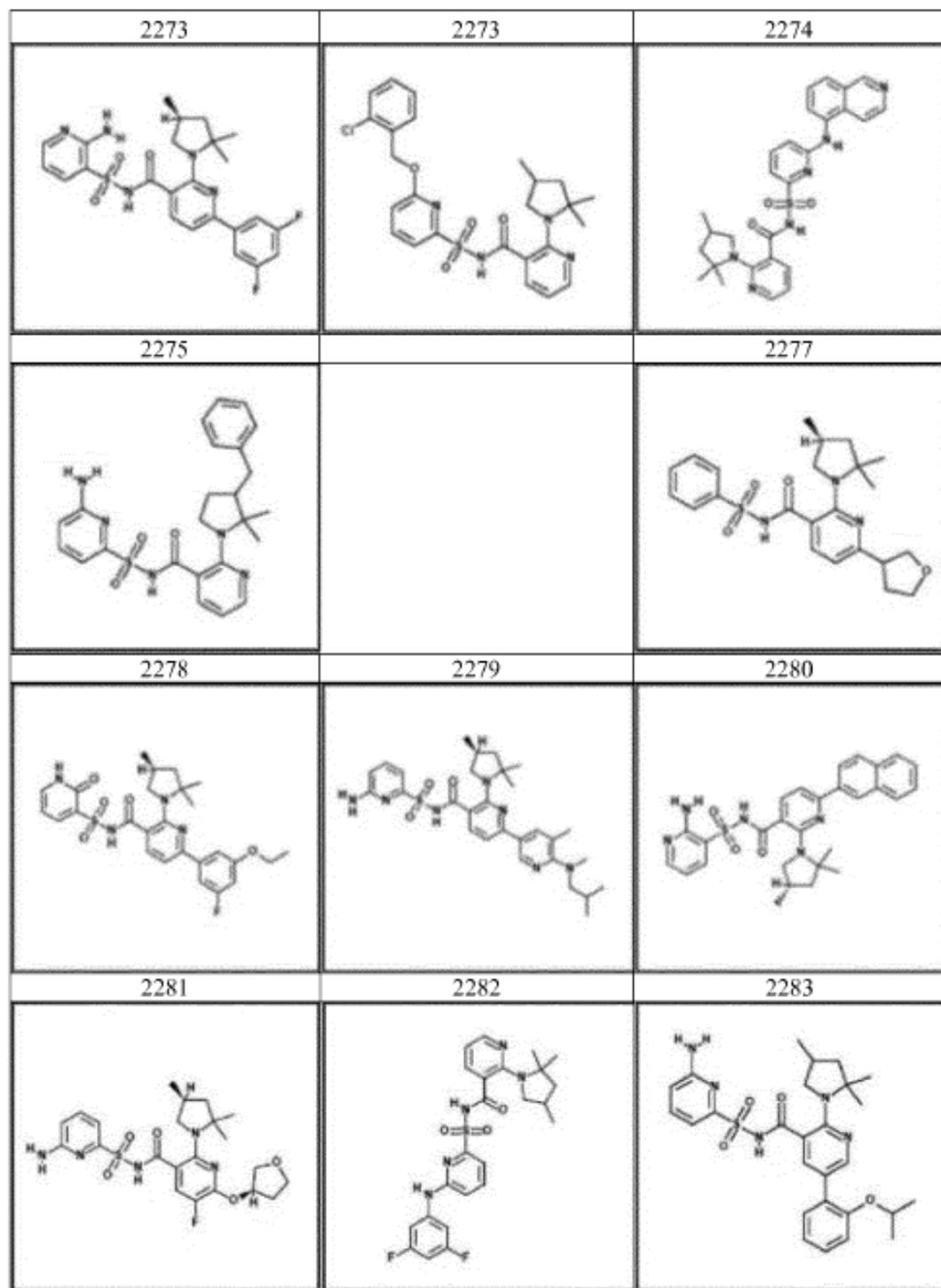




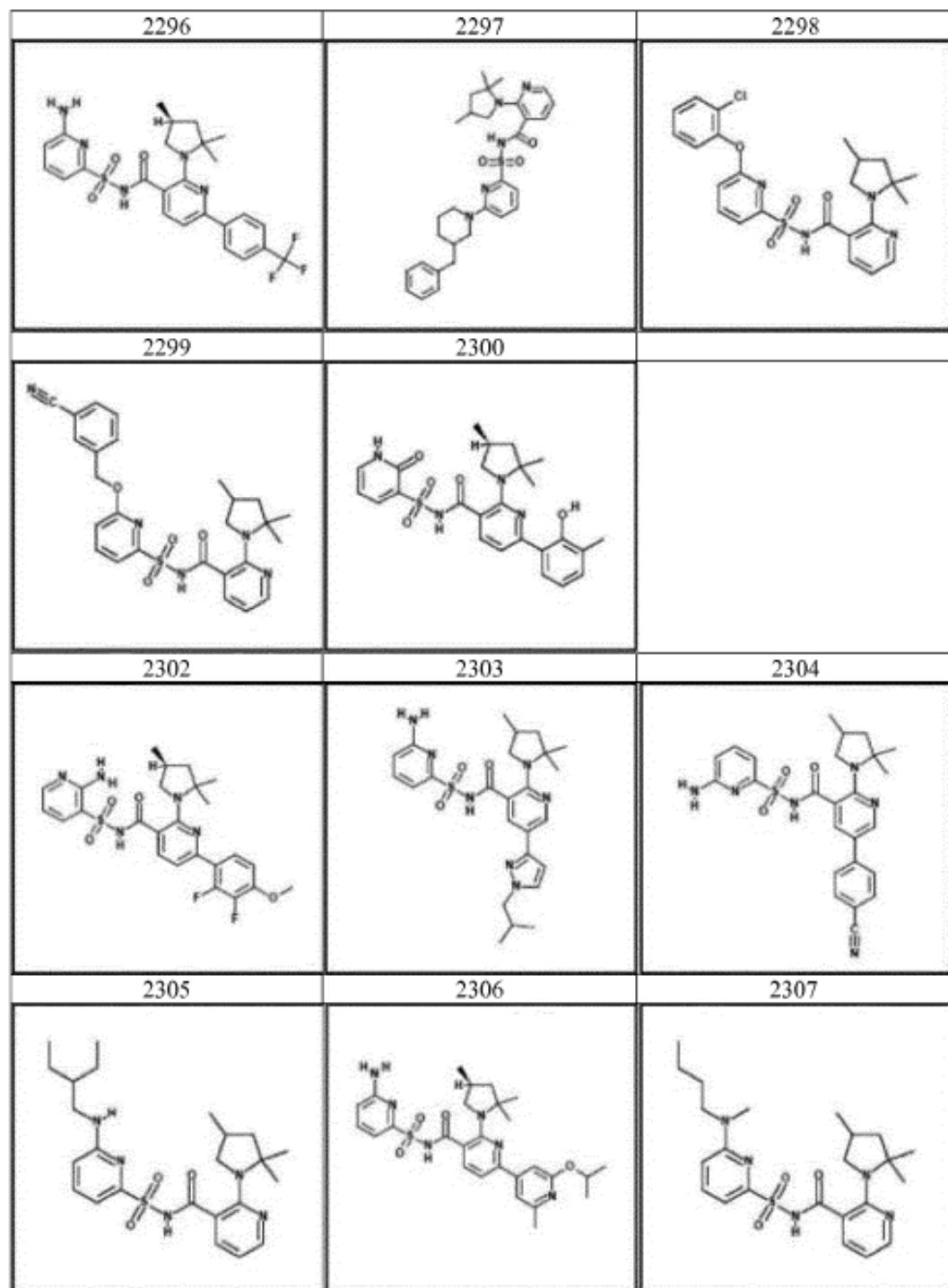


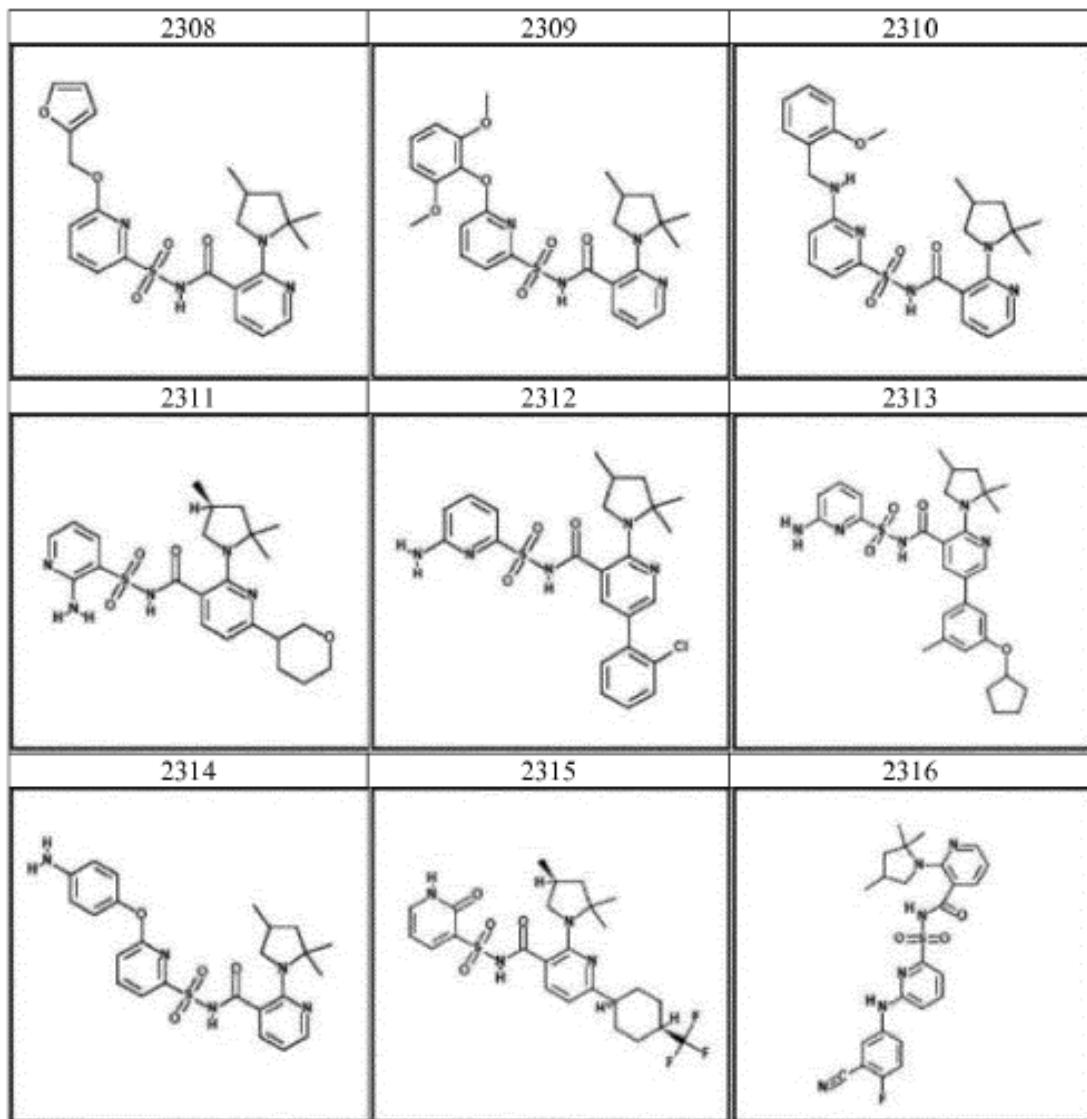


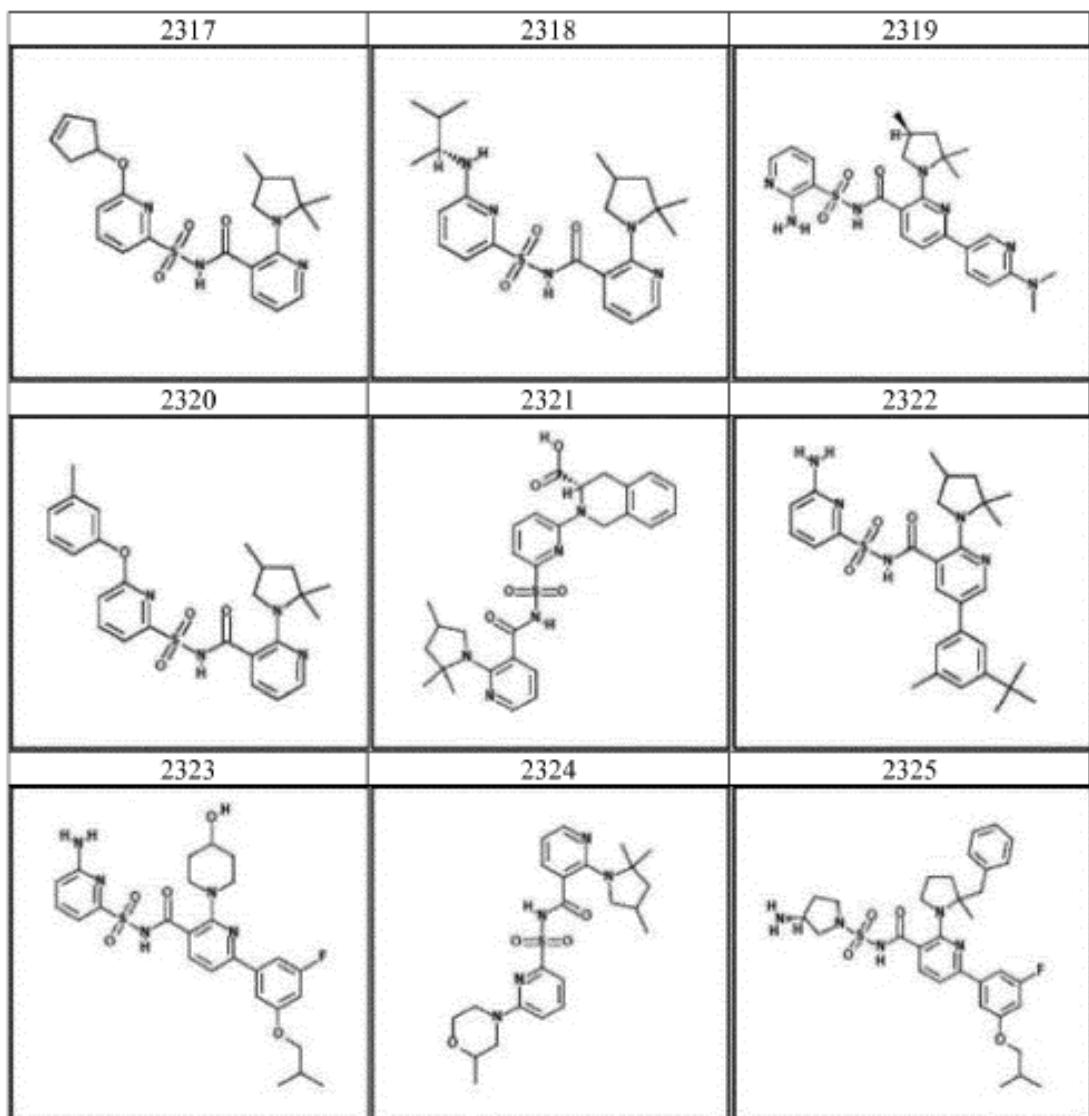


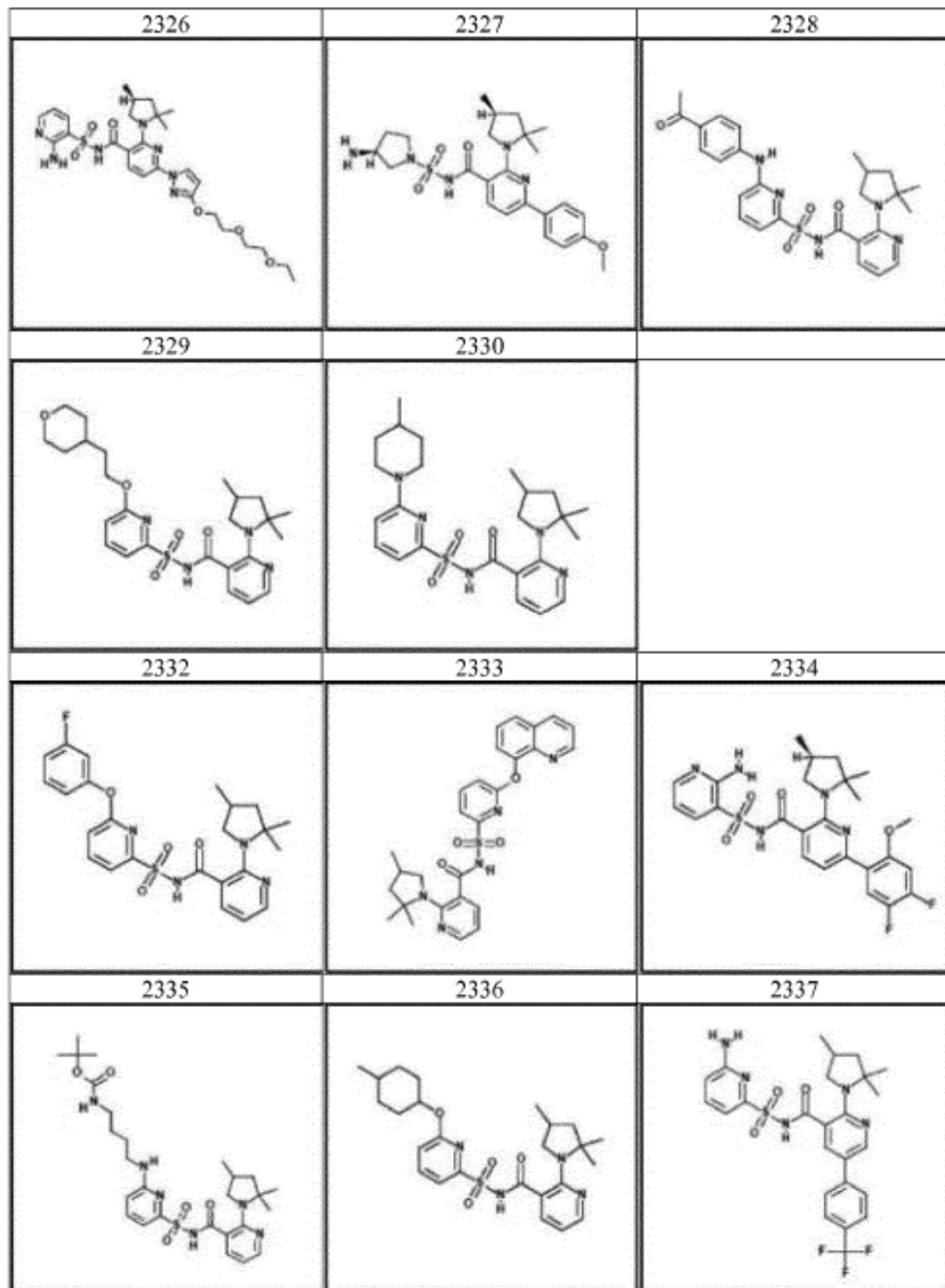


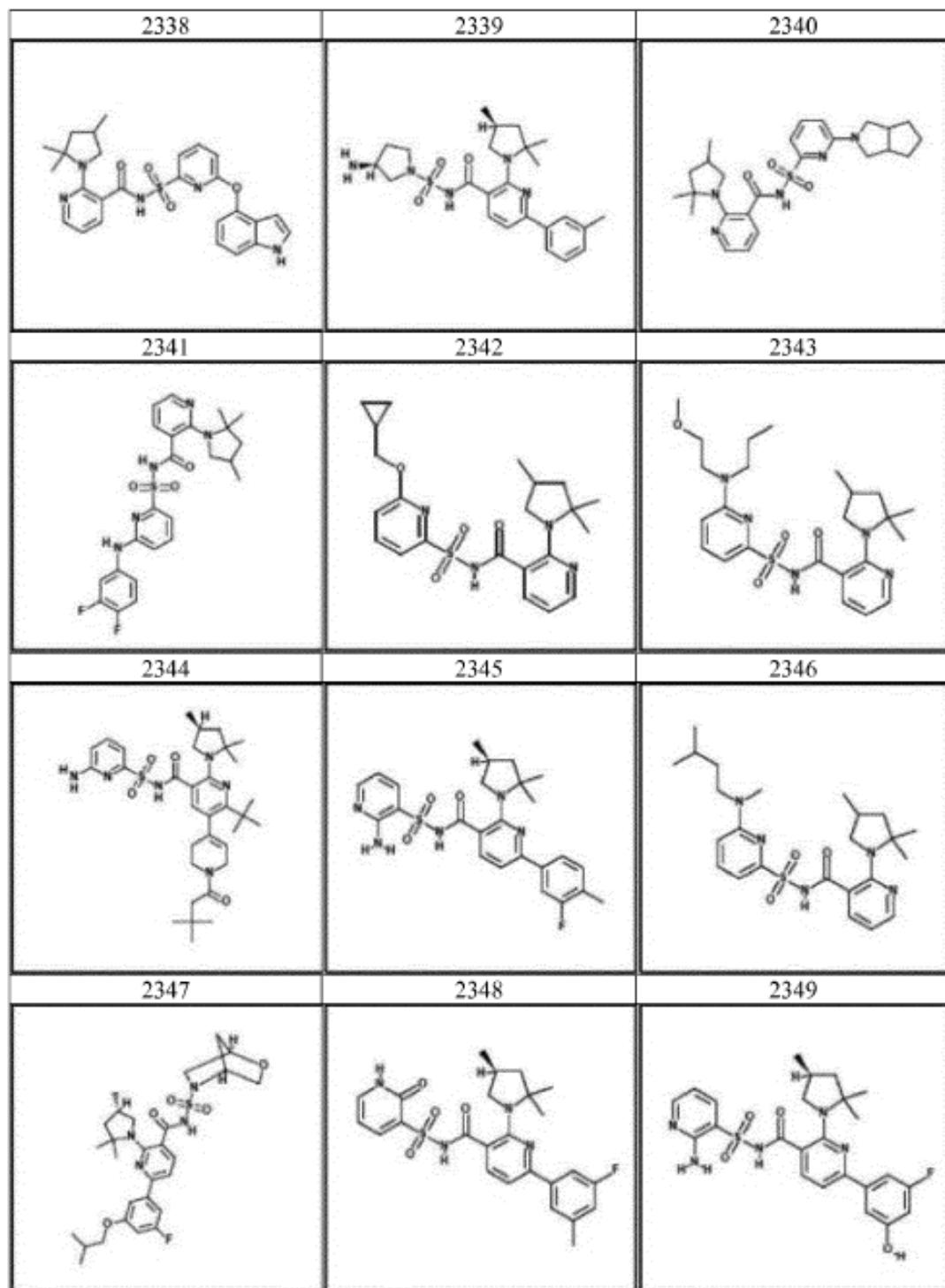
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2293	2294	2295

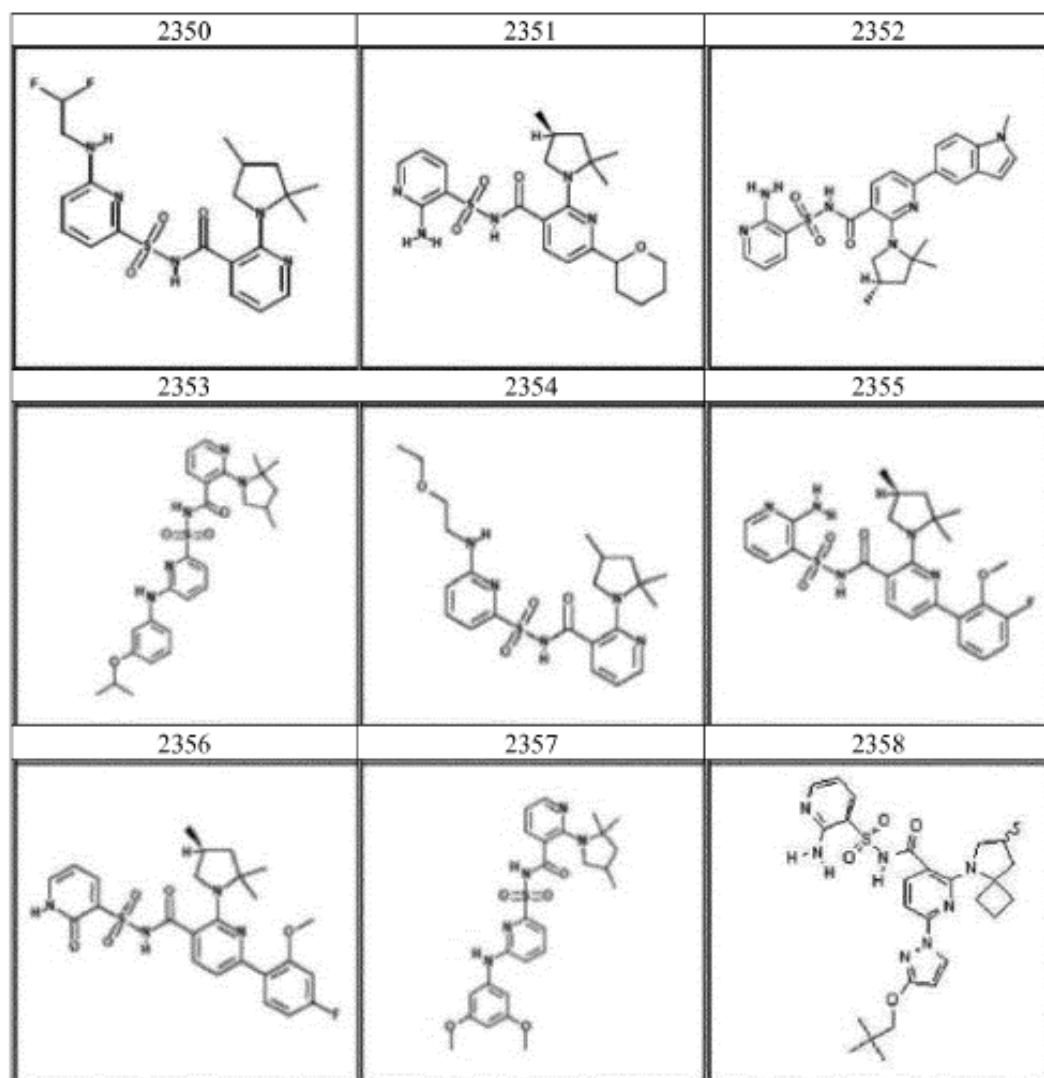


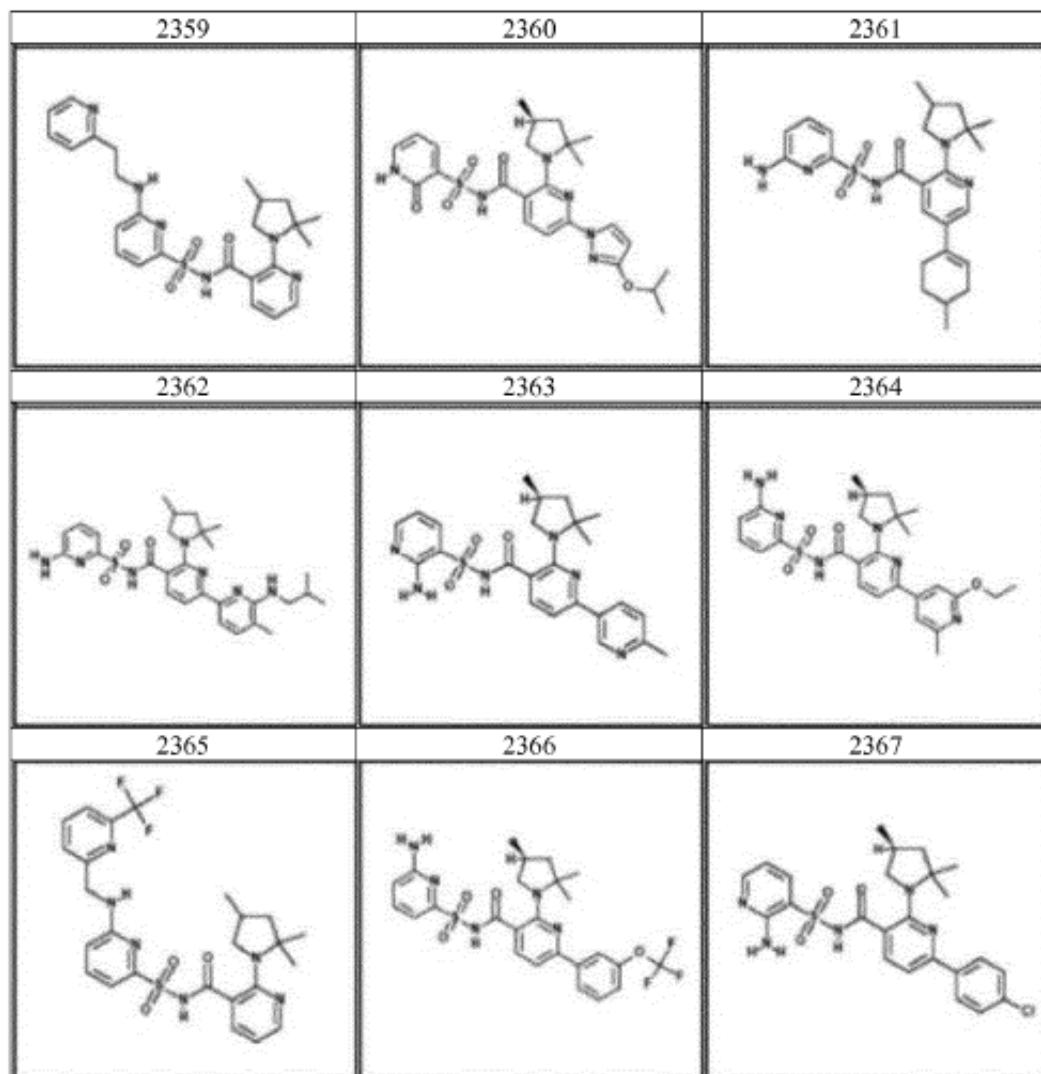


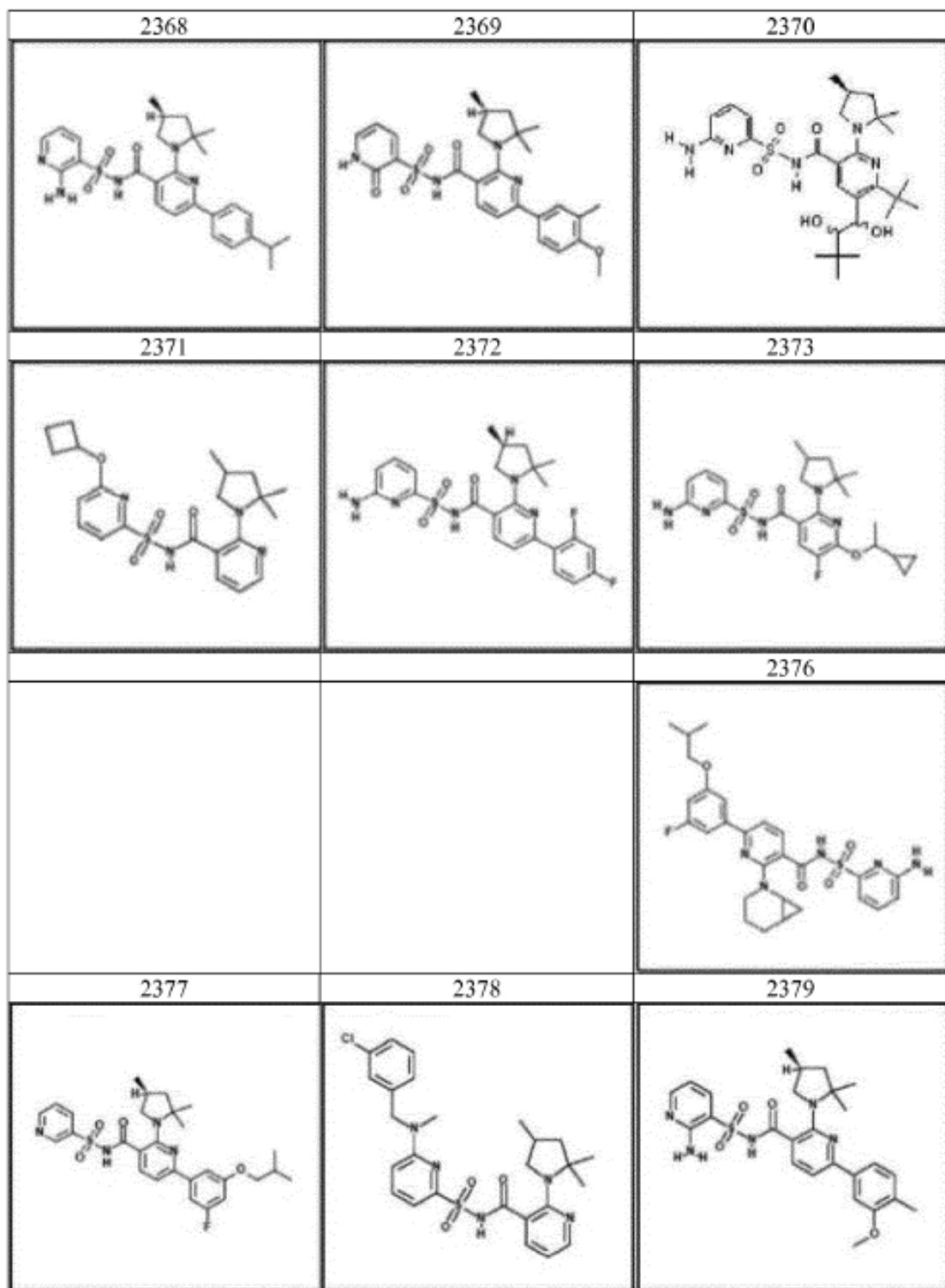


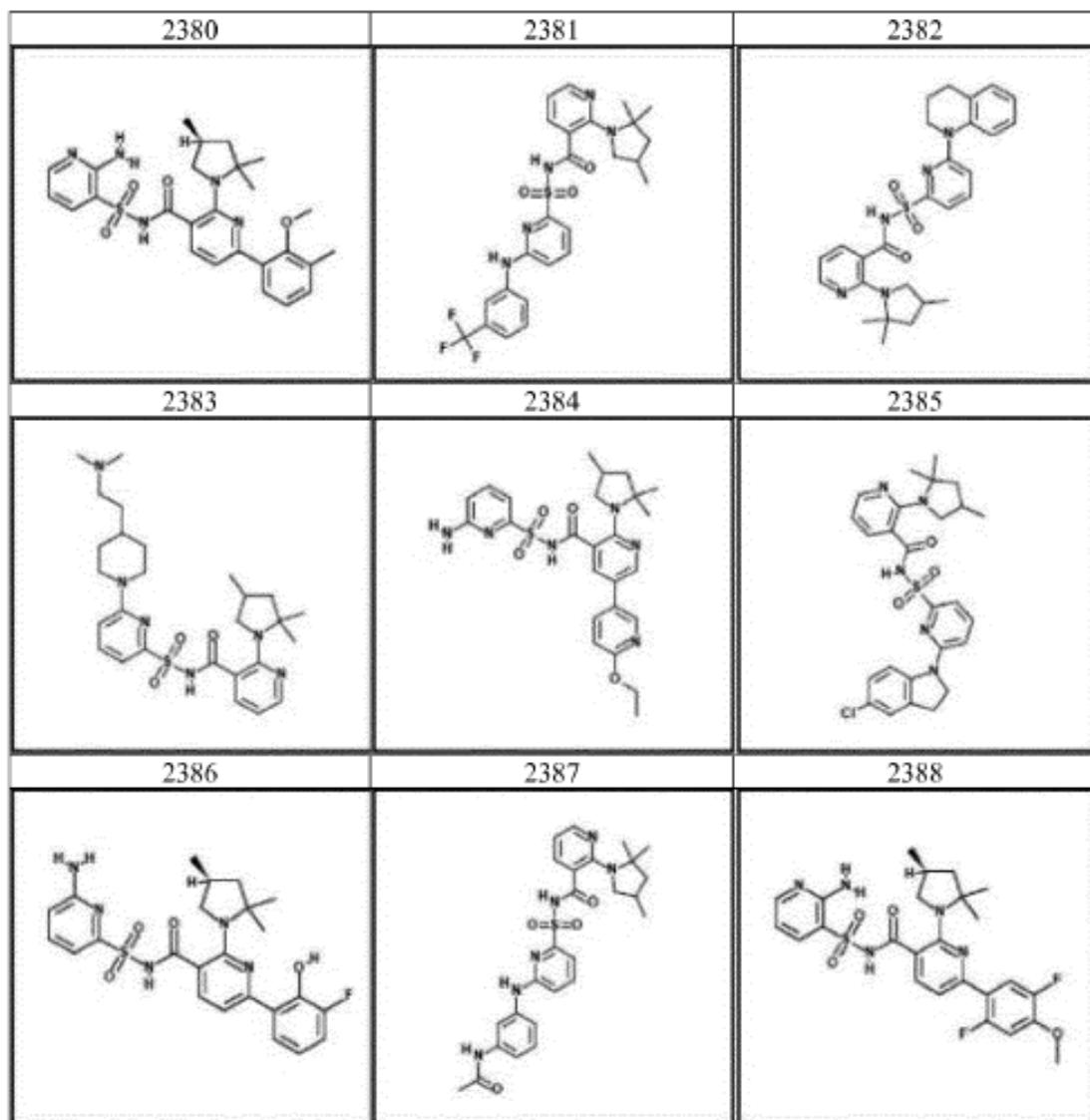


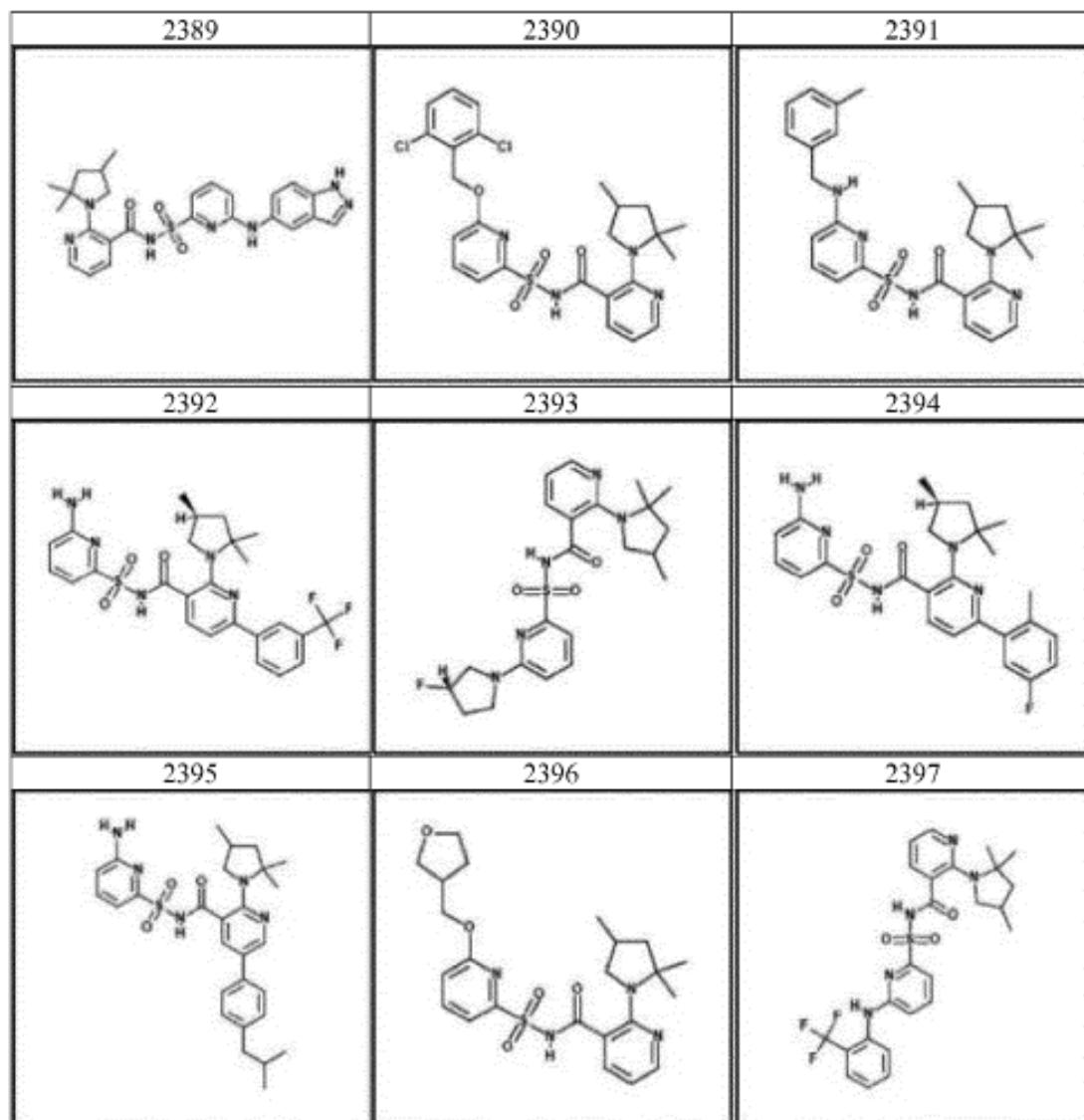


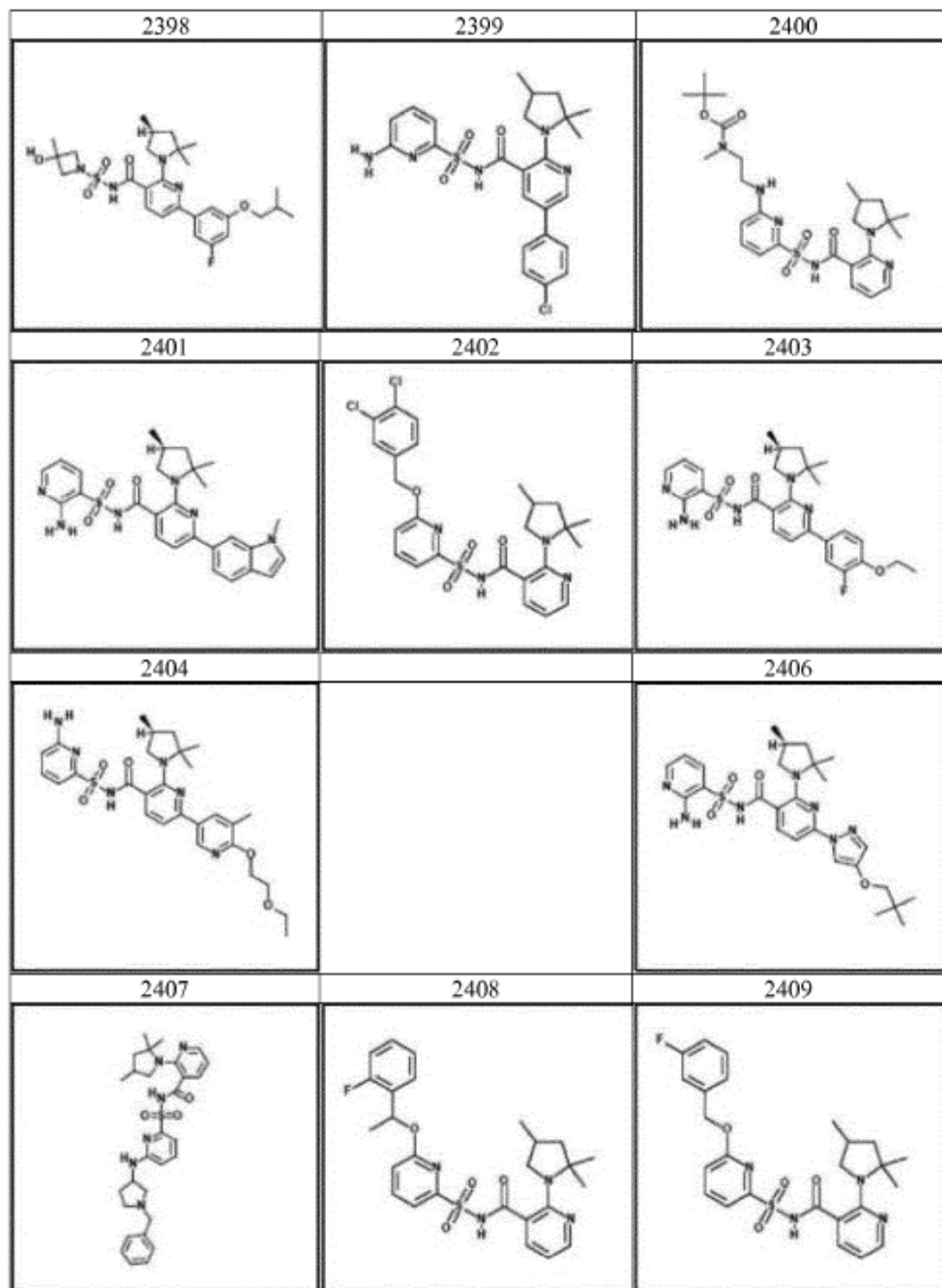


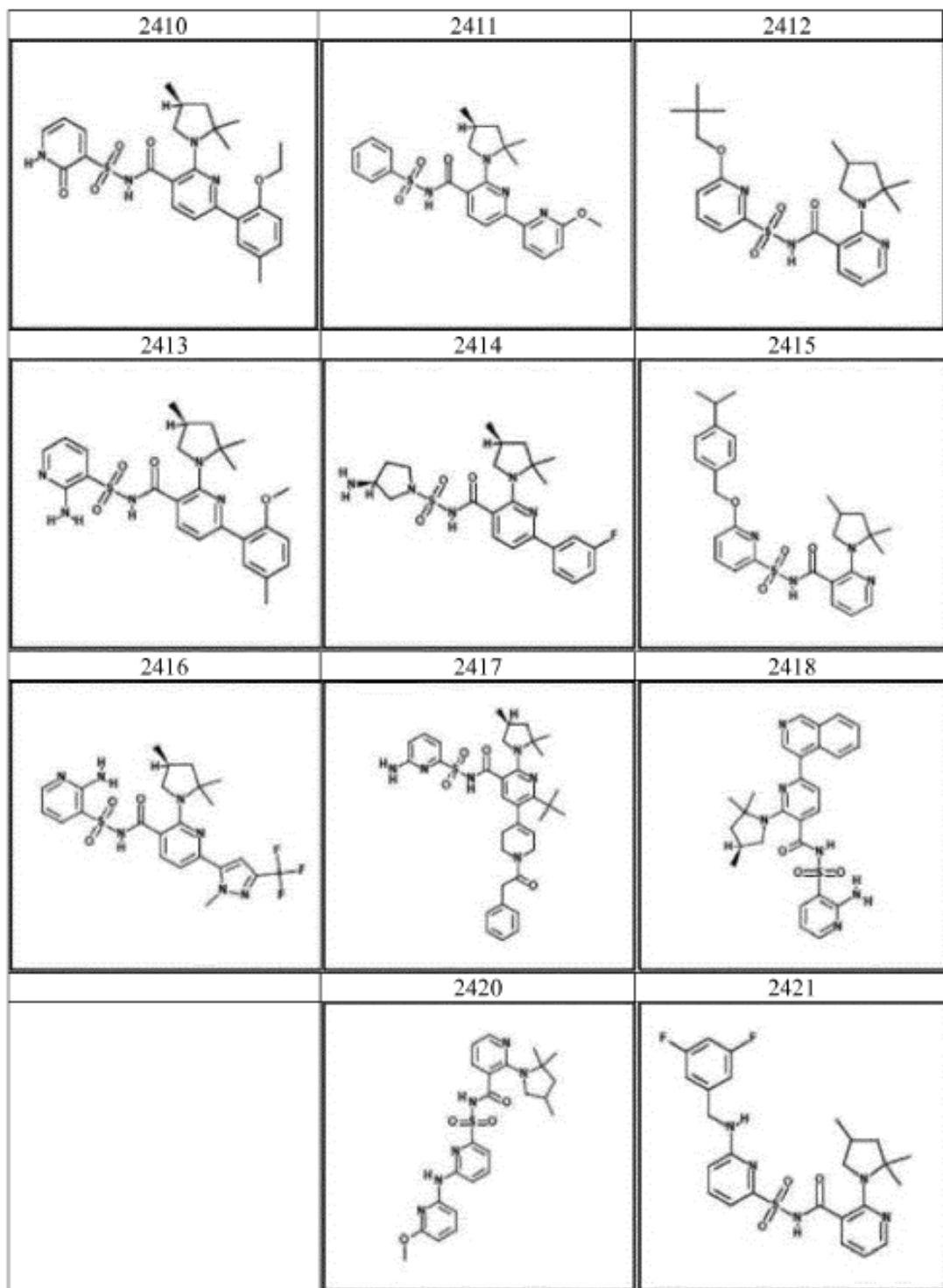


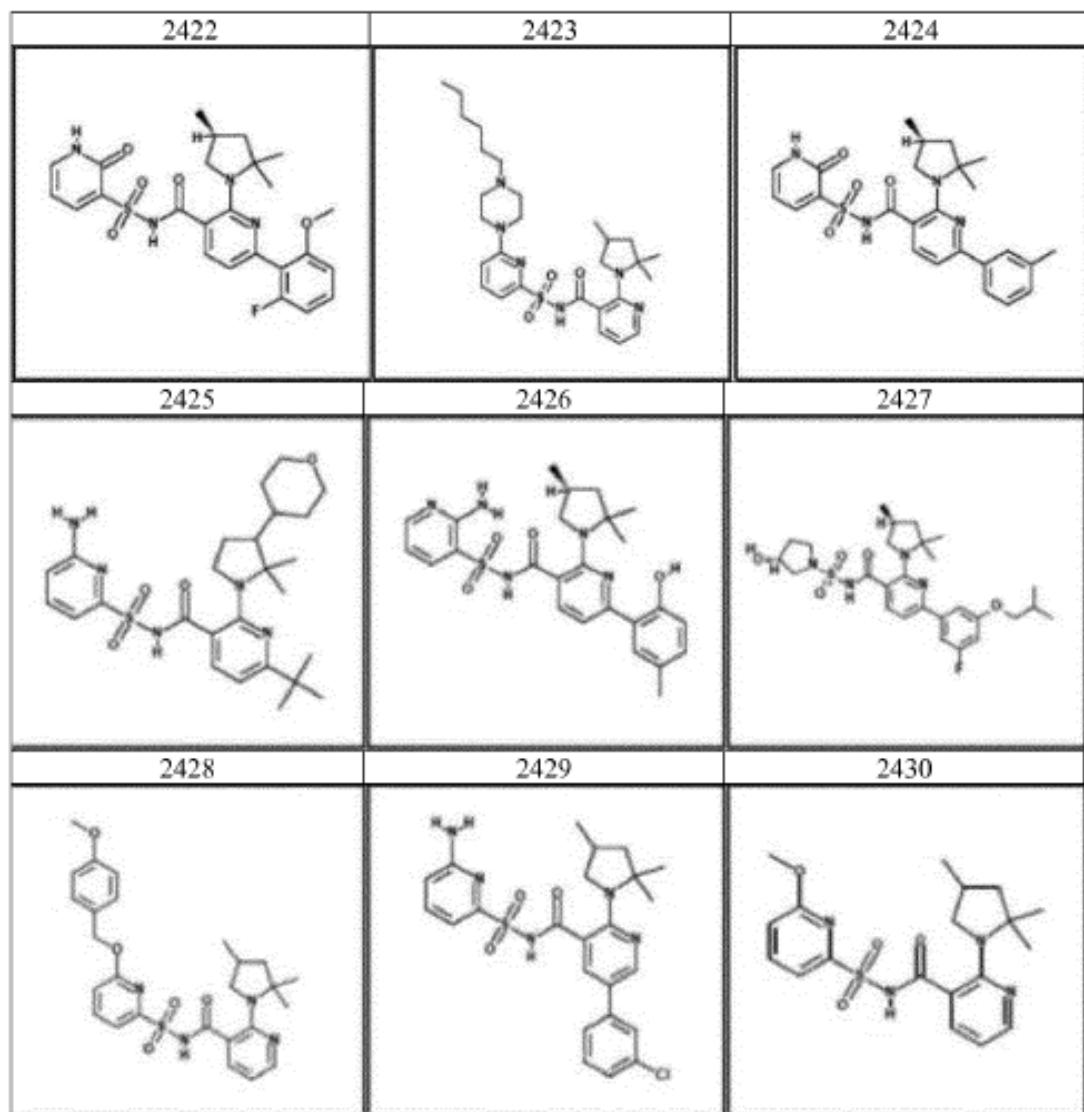


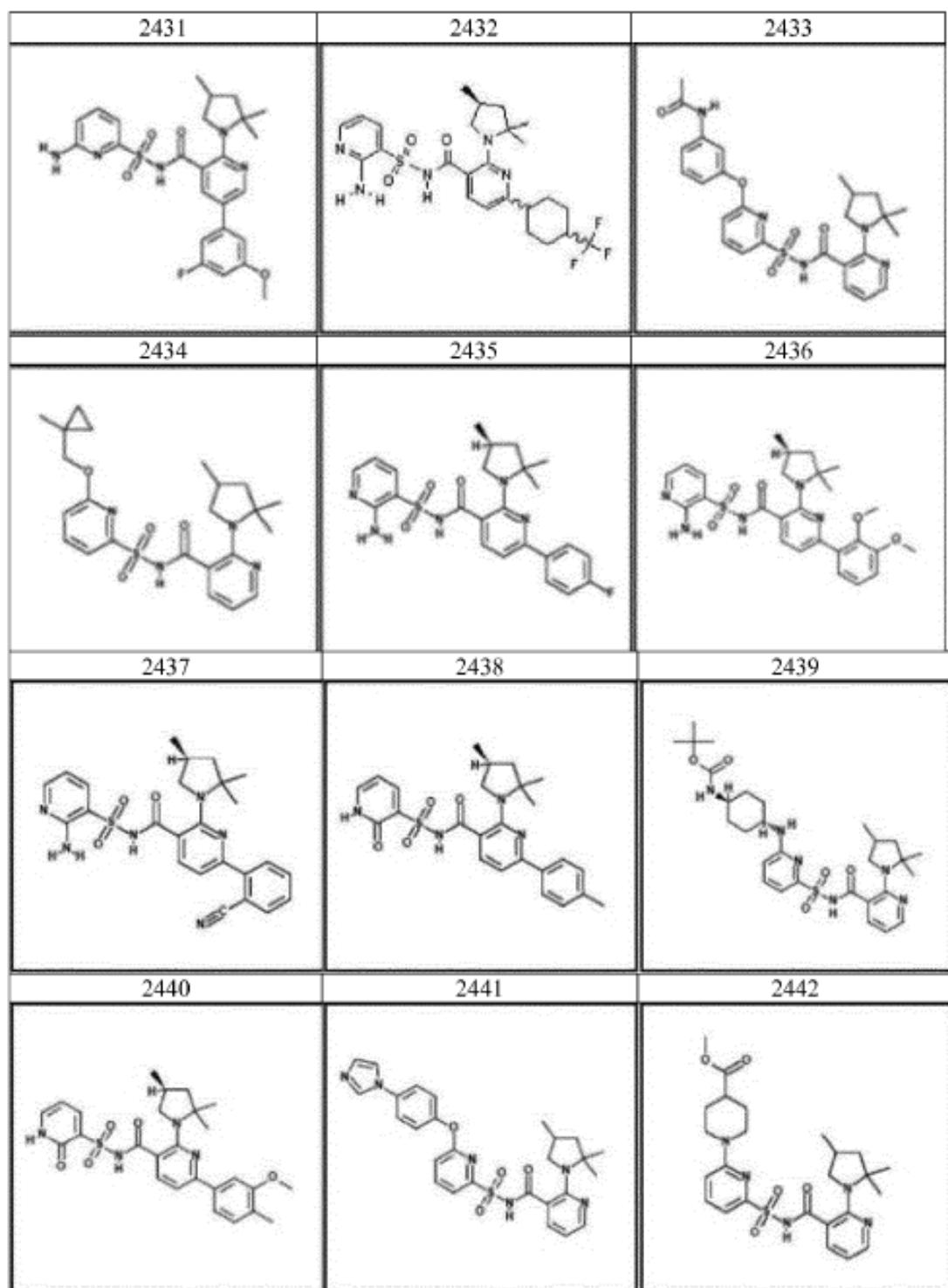


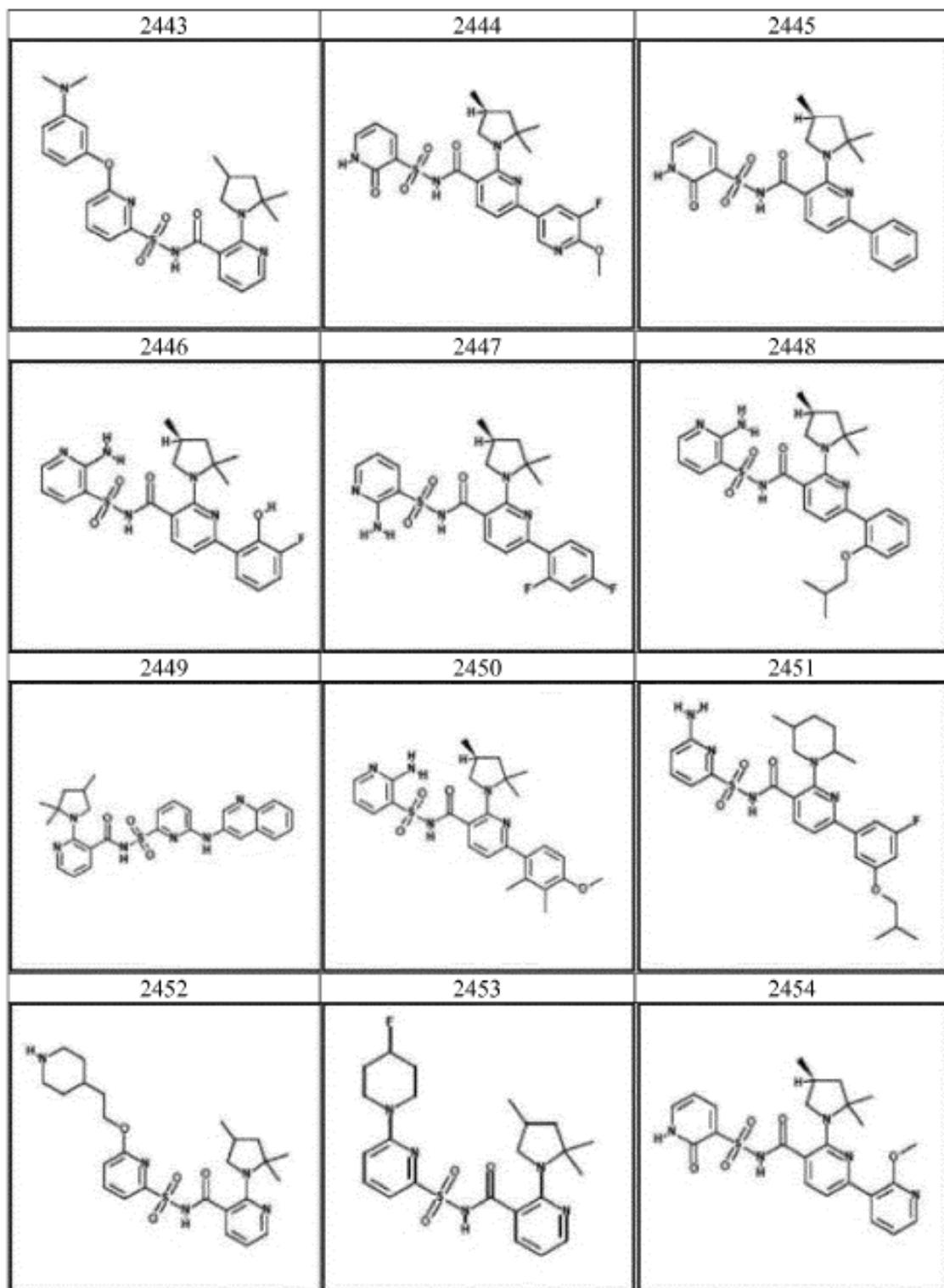


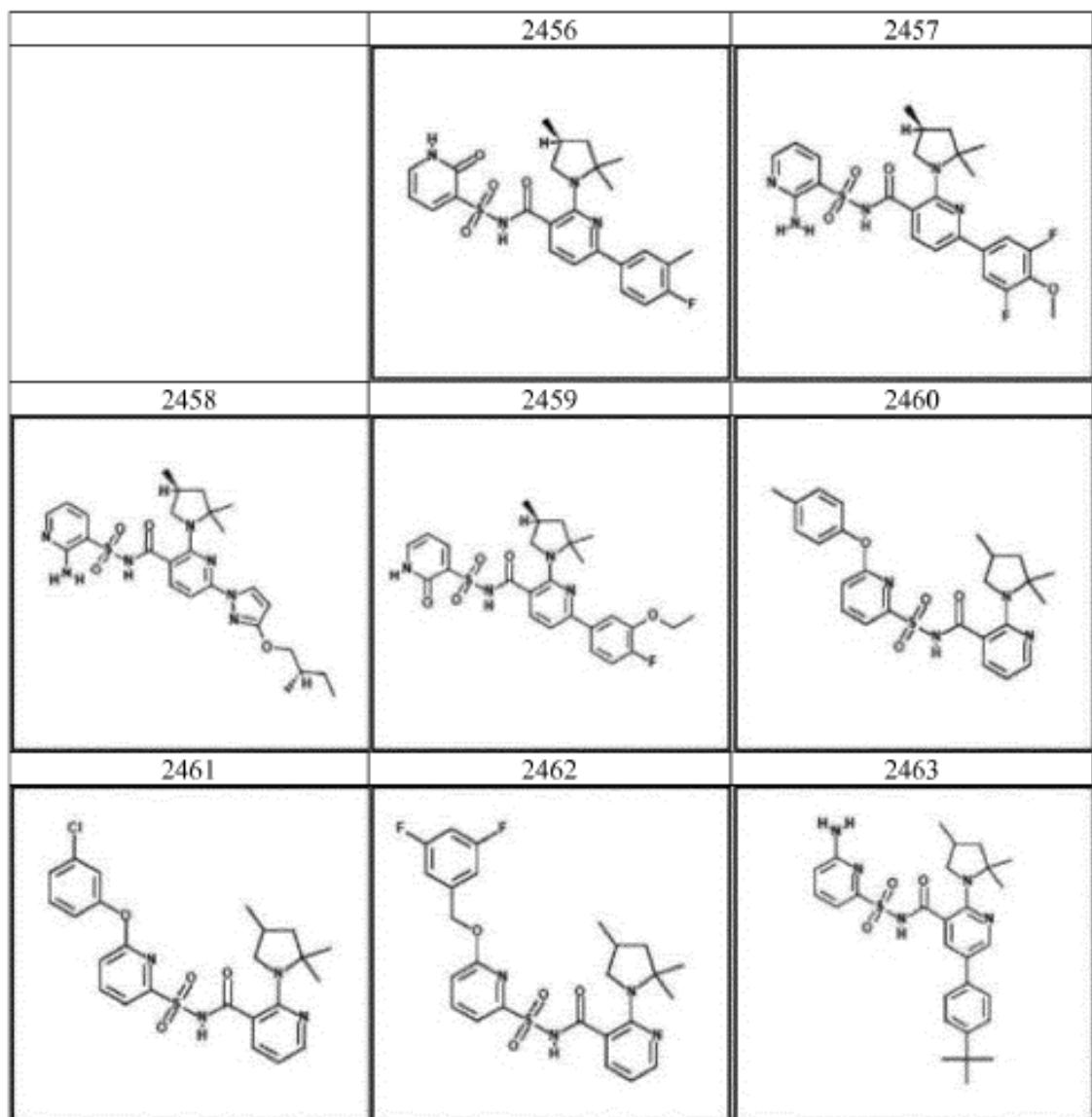


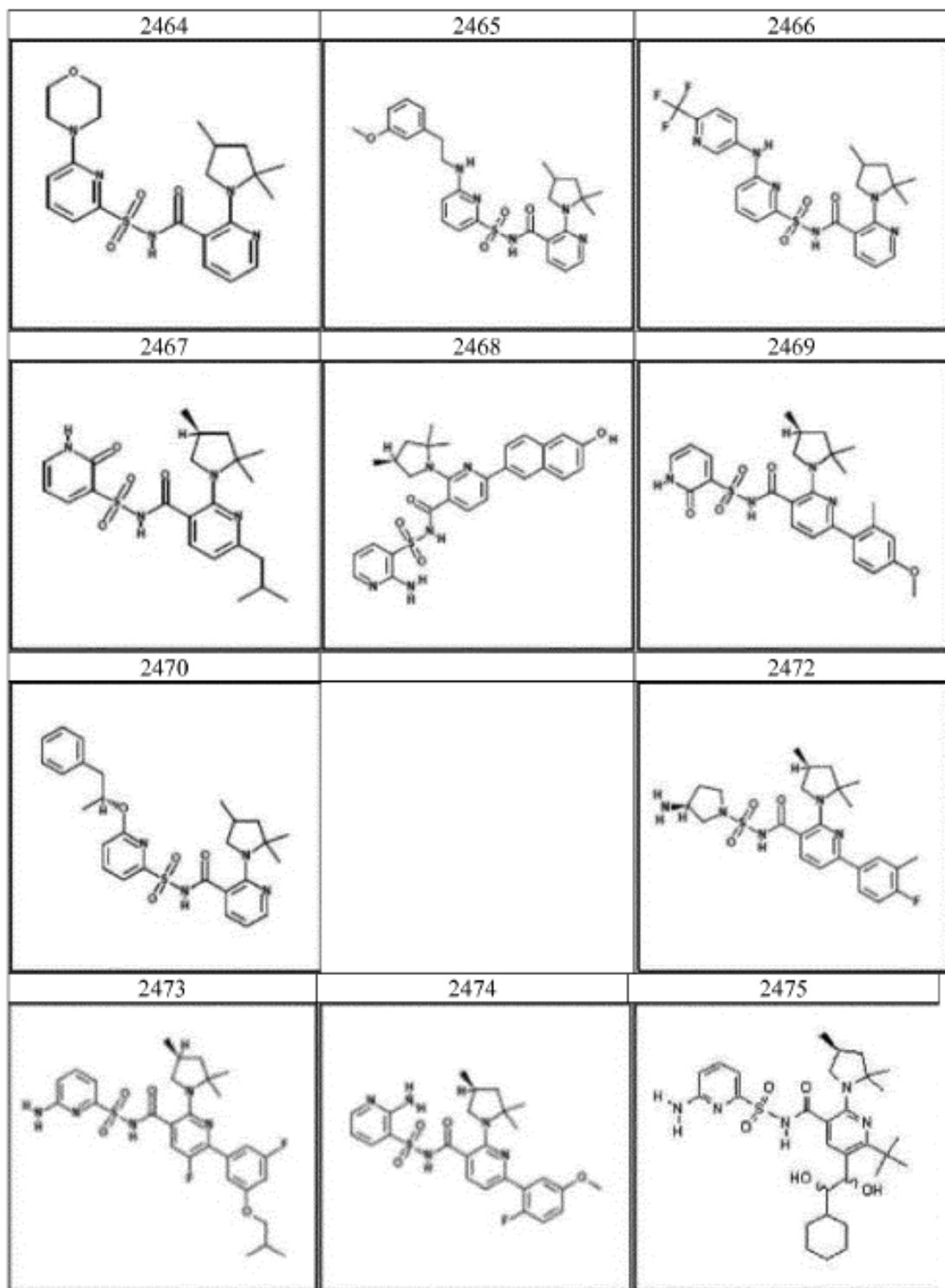


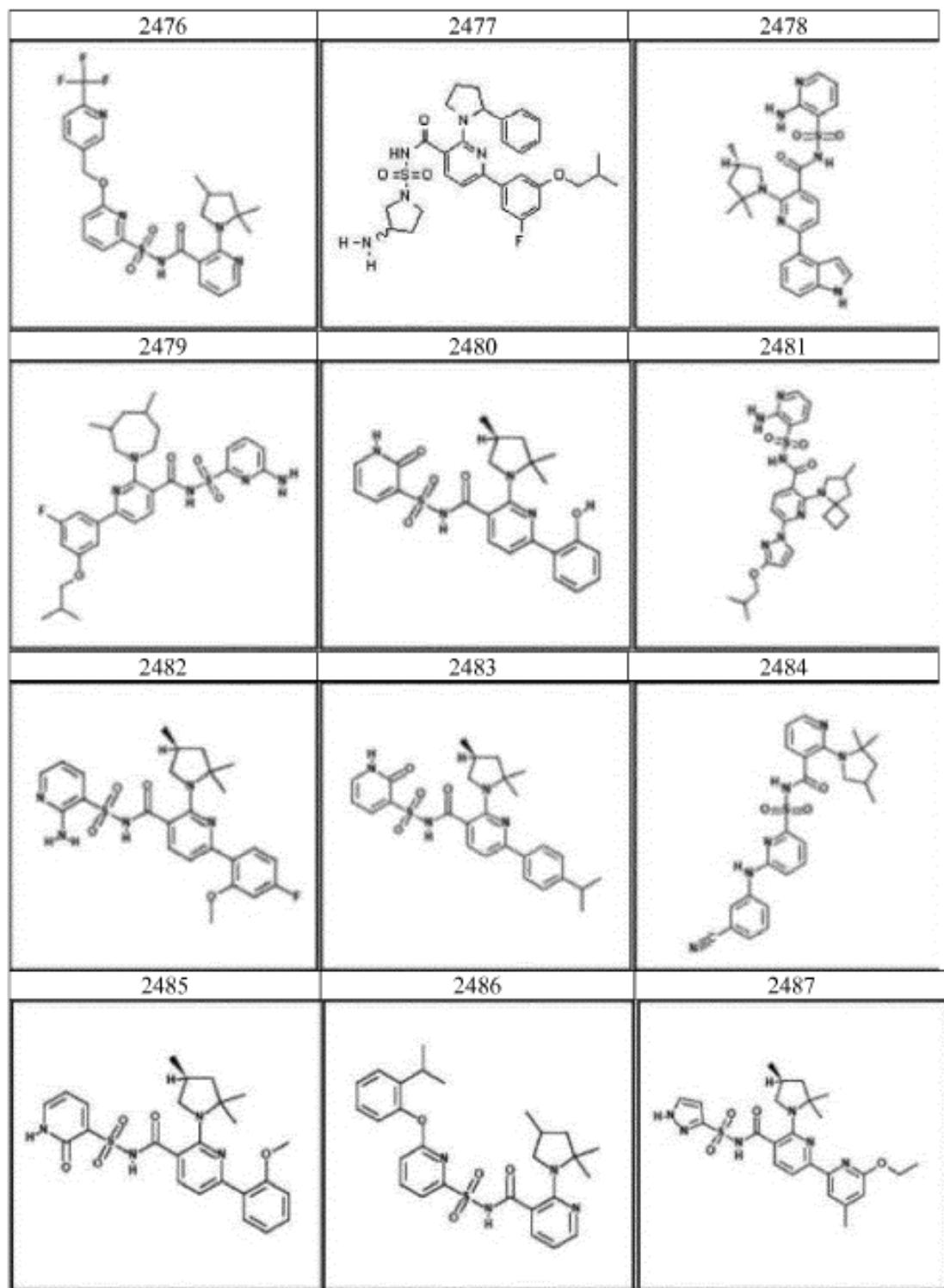


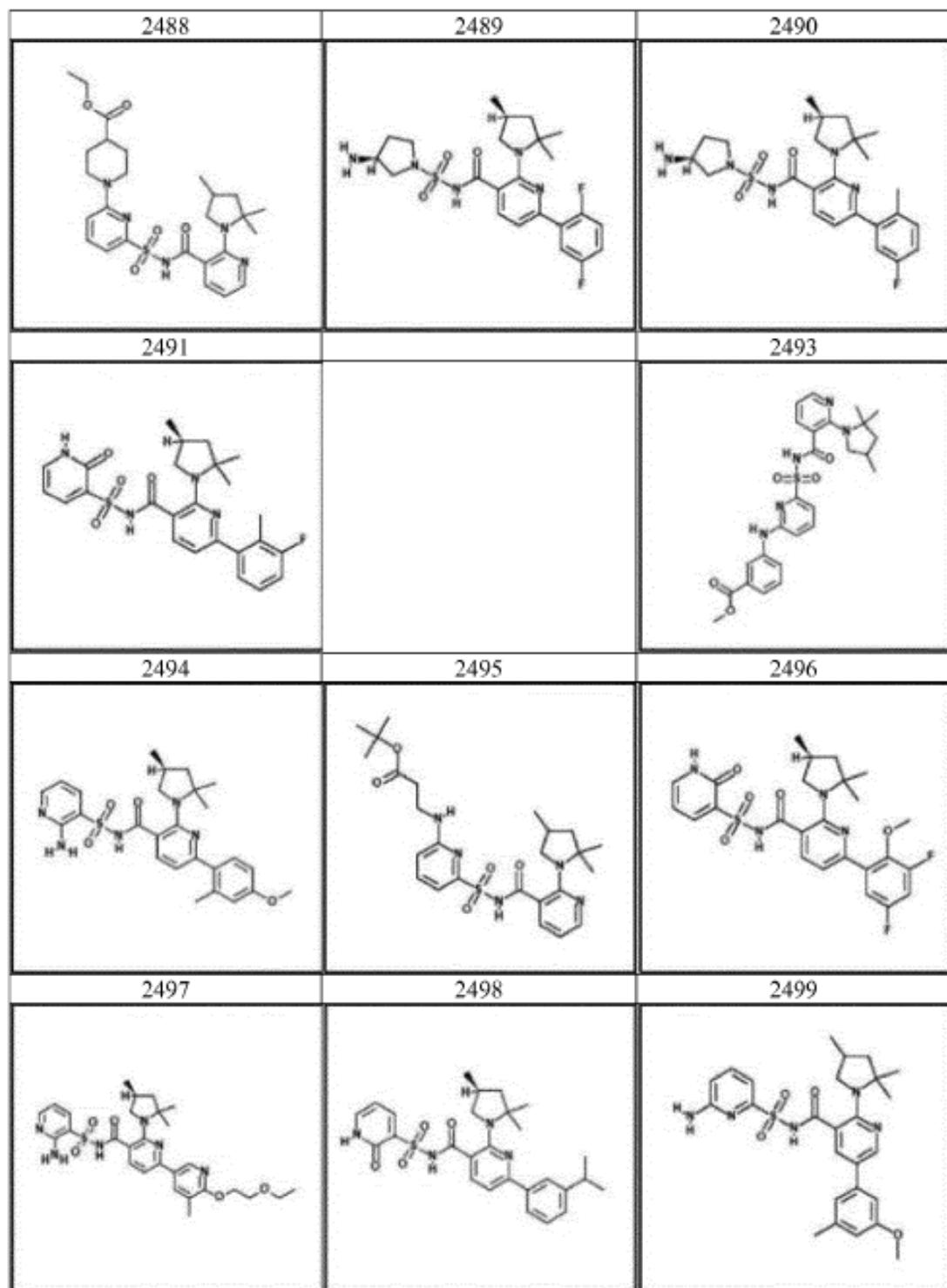


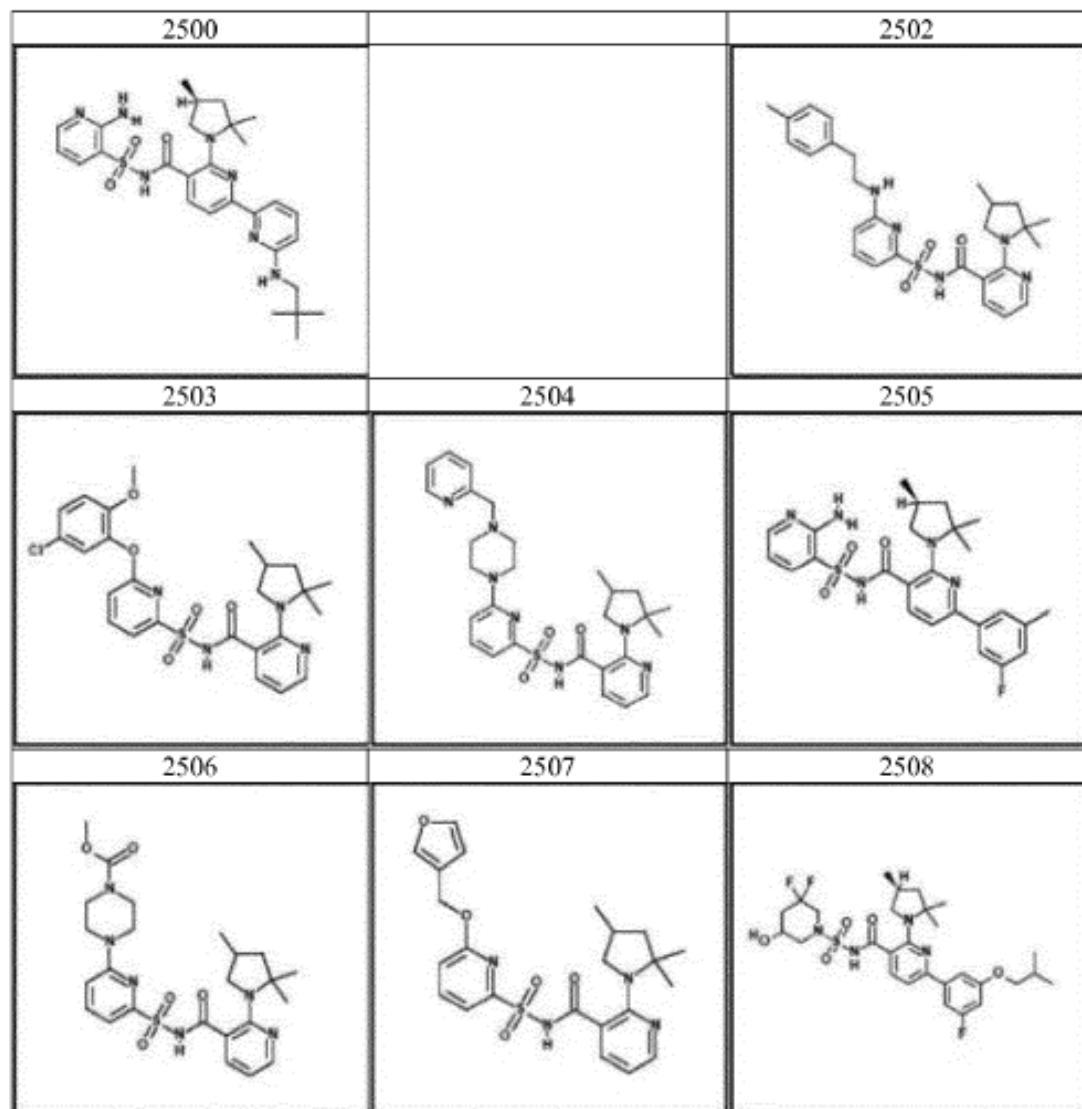


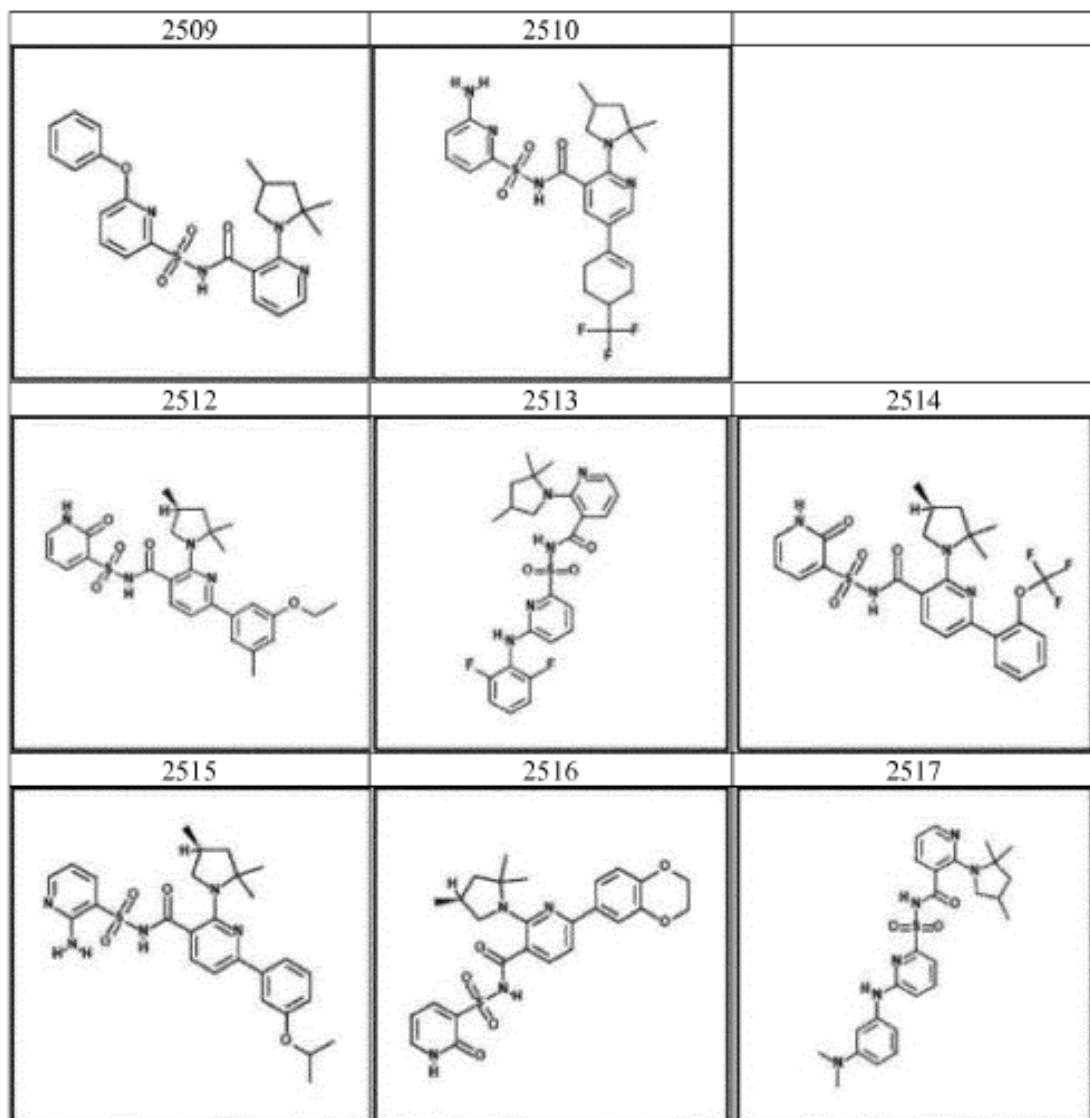


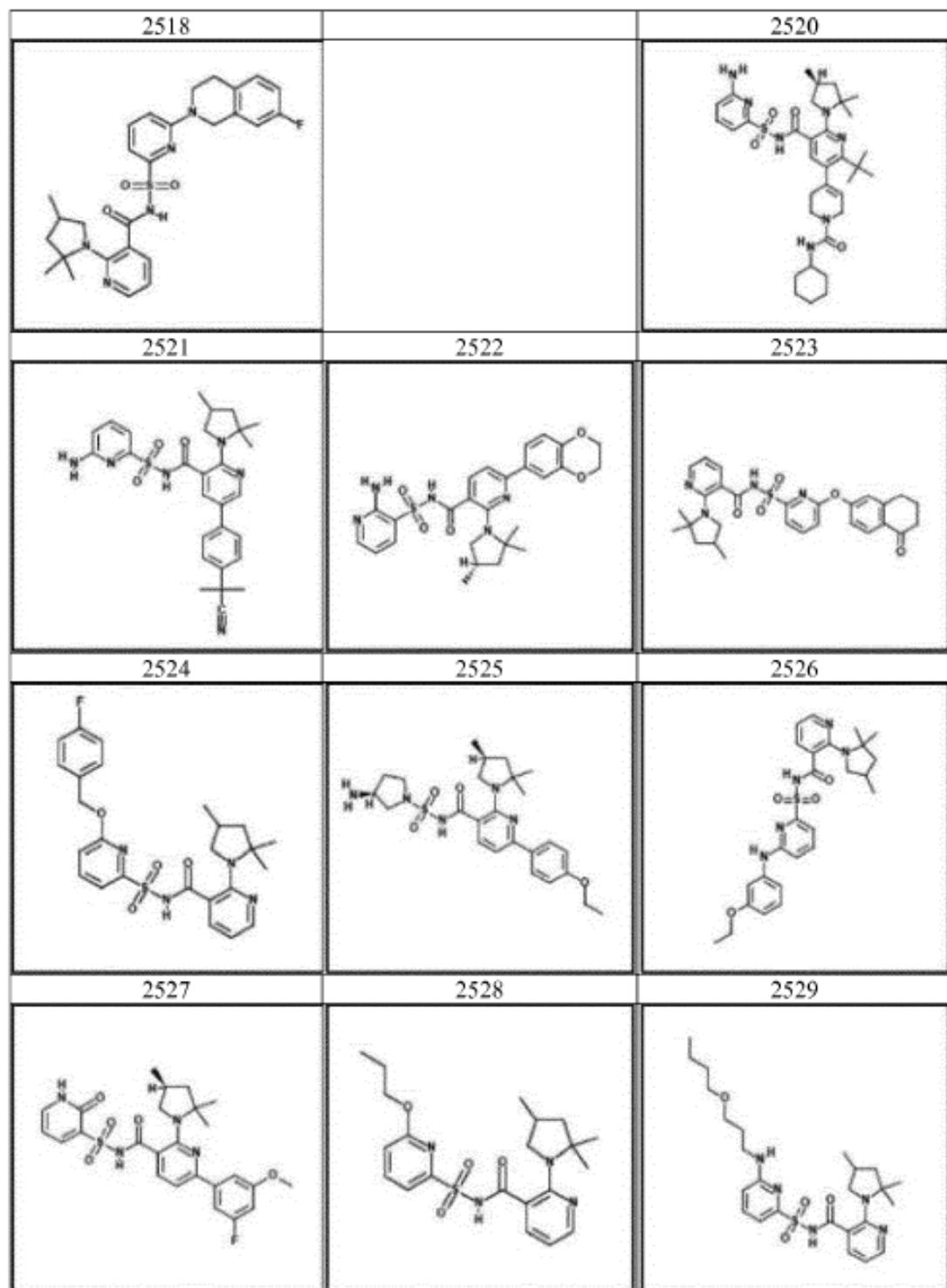


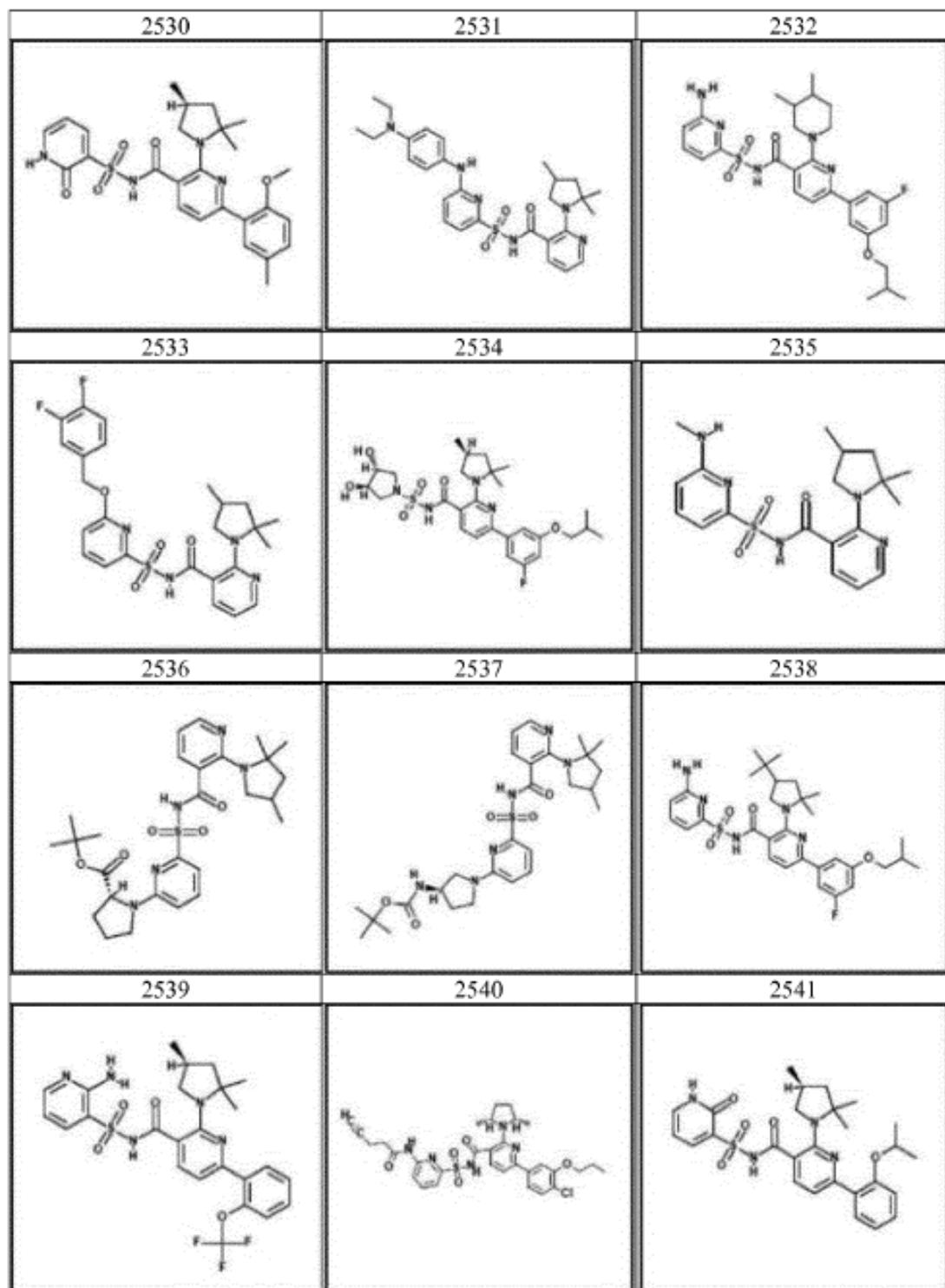




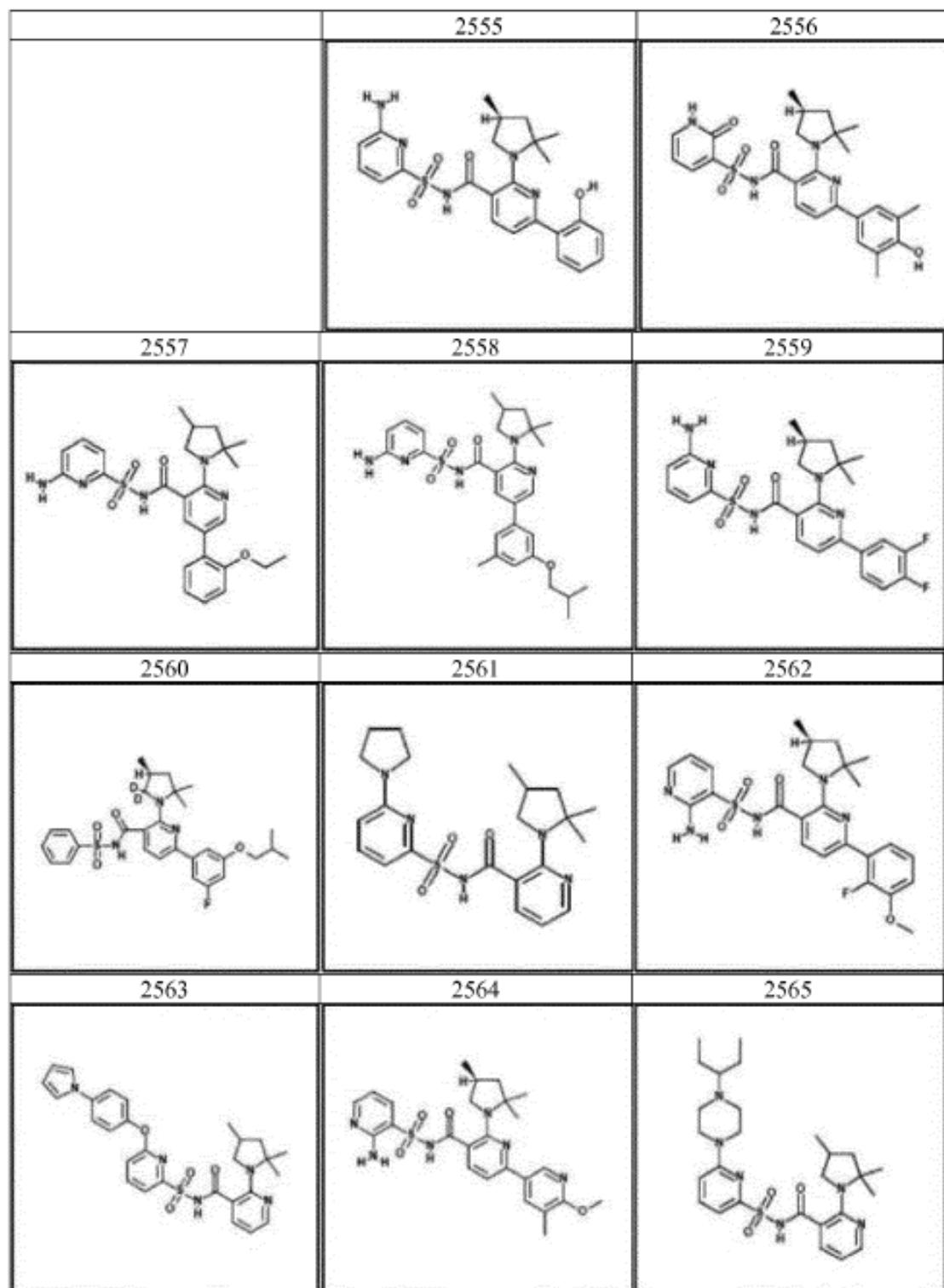


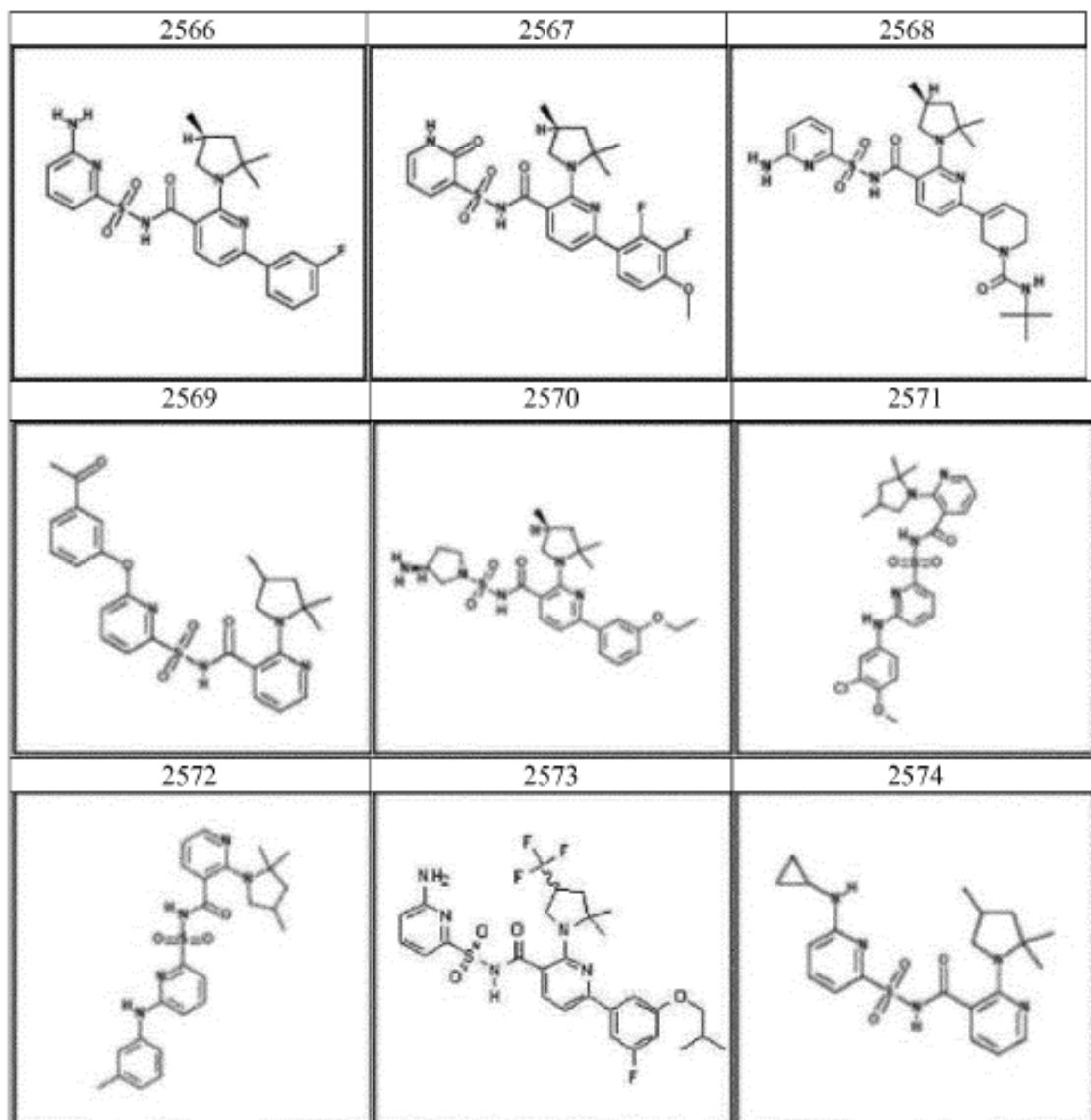


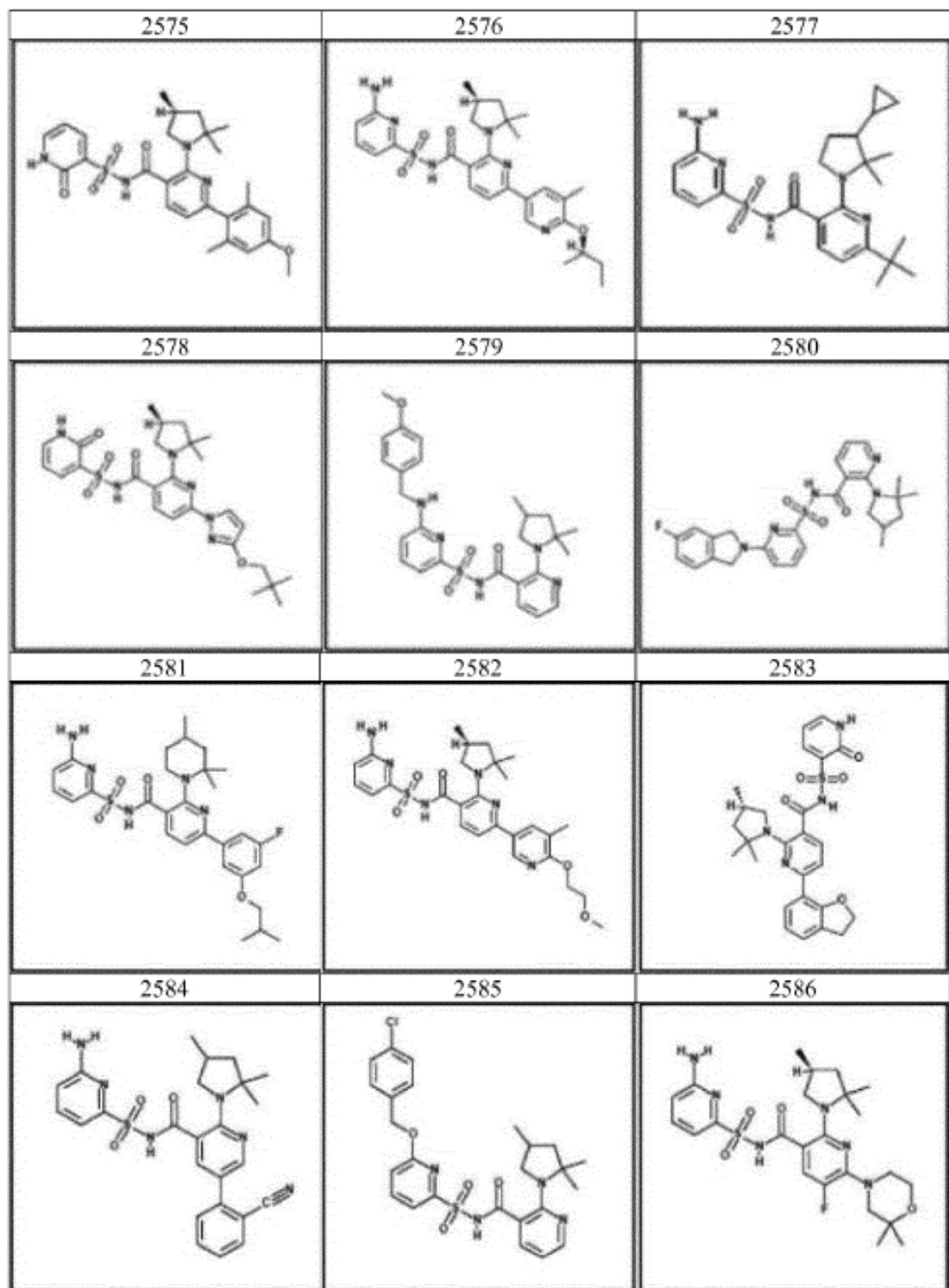


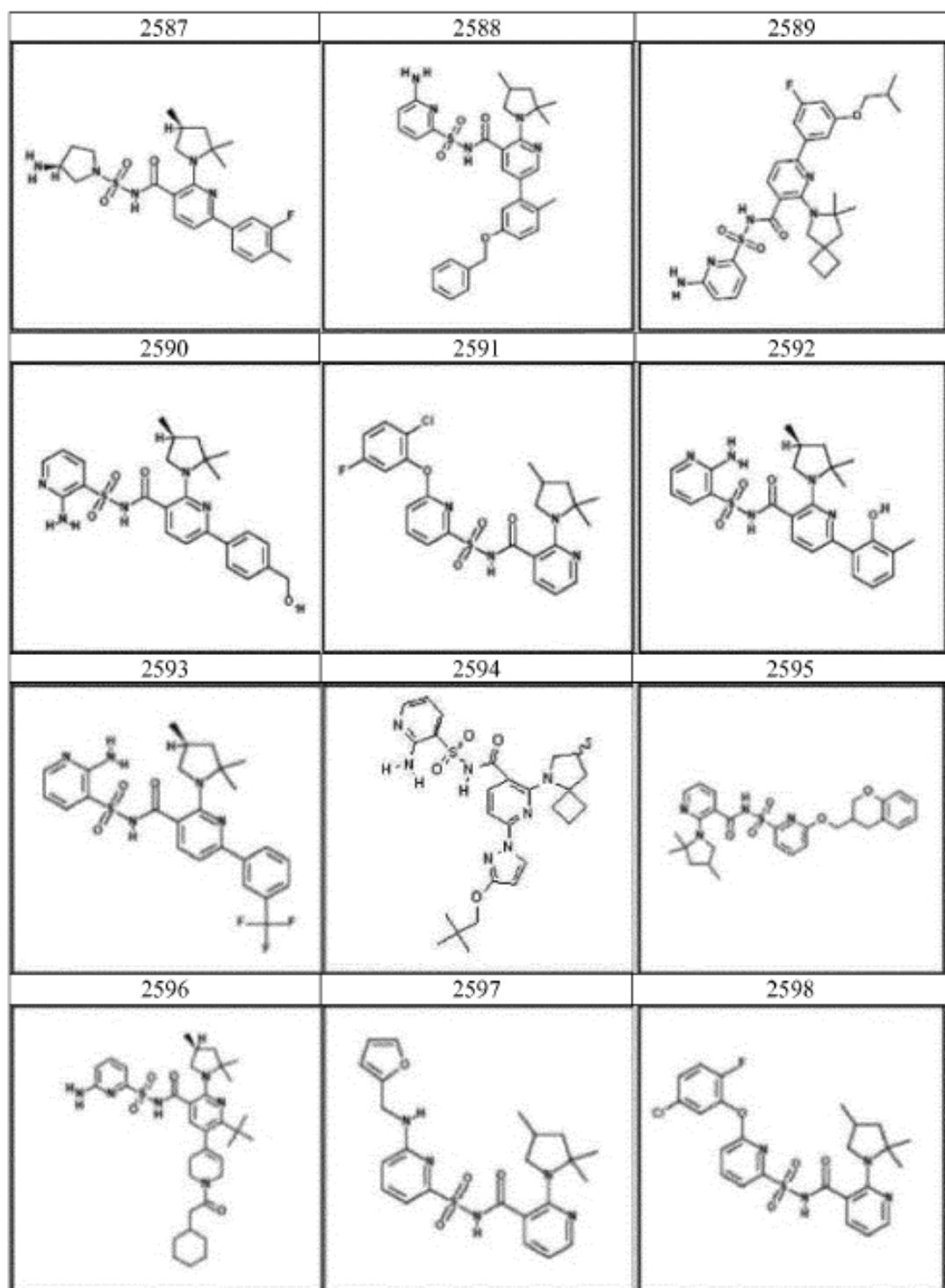


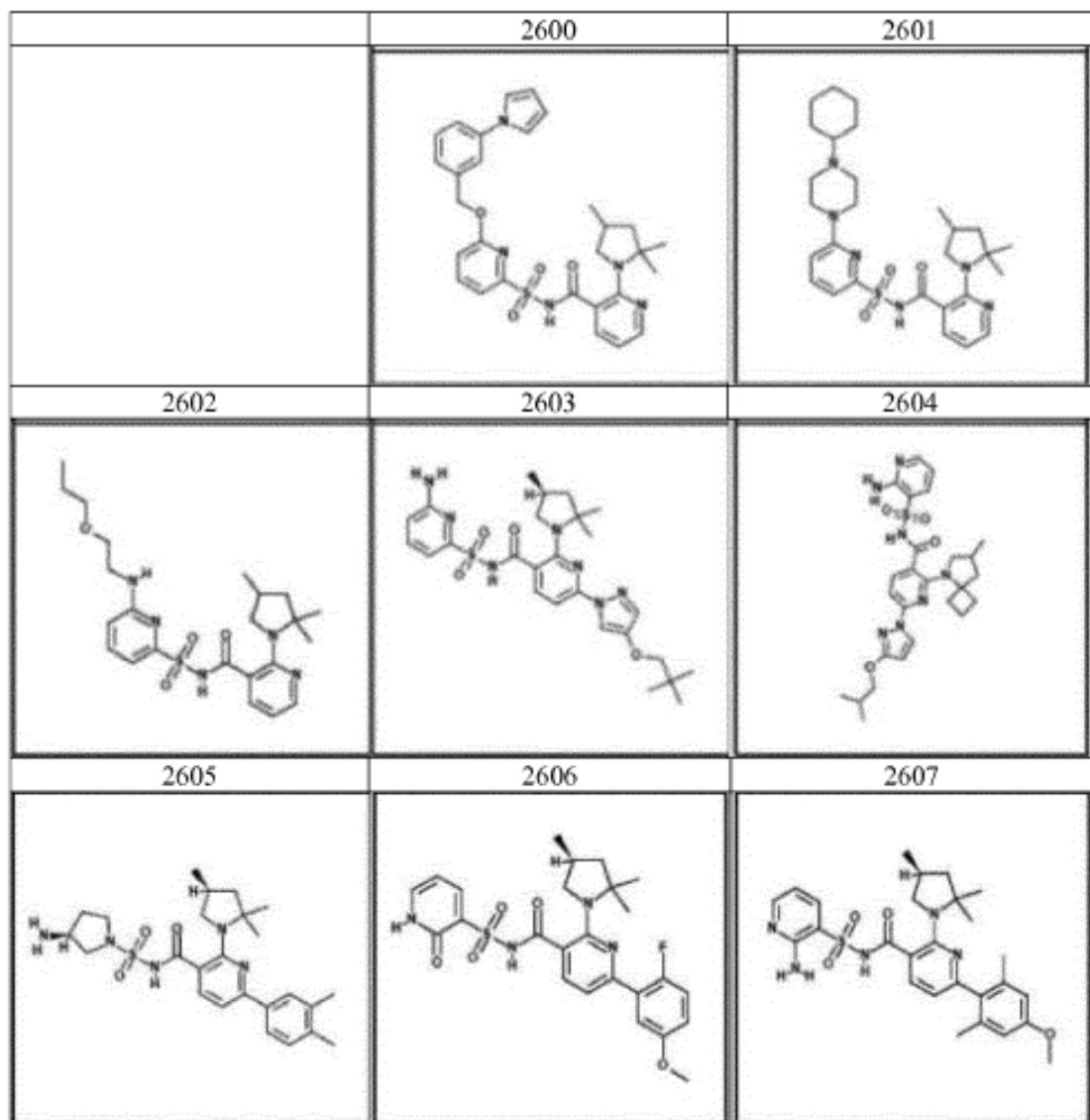
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2551	2552	2553

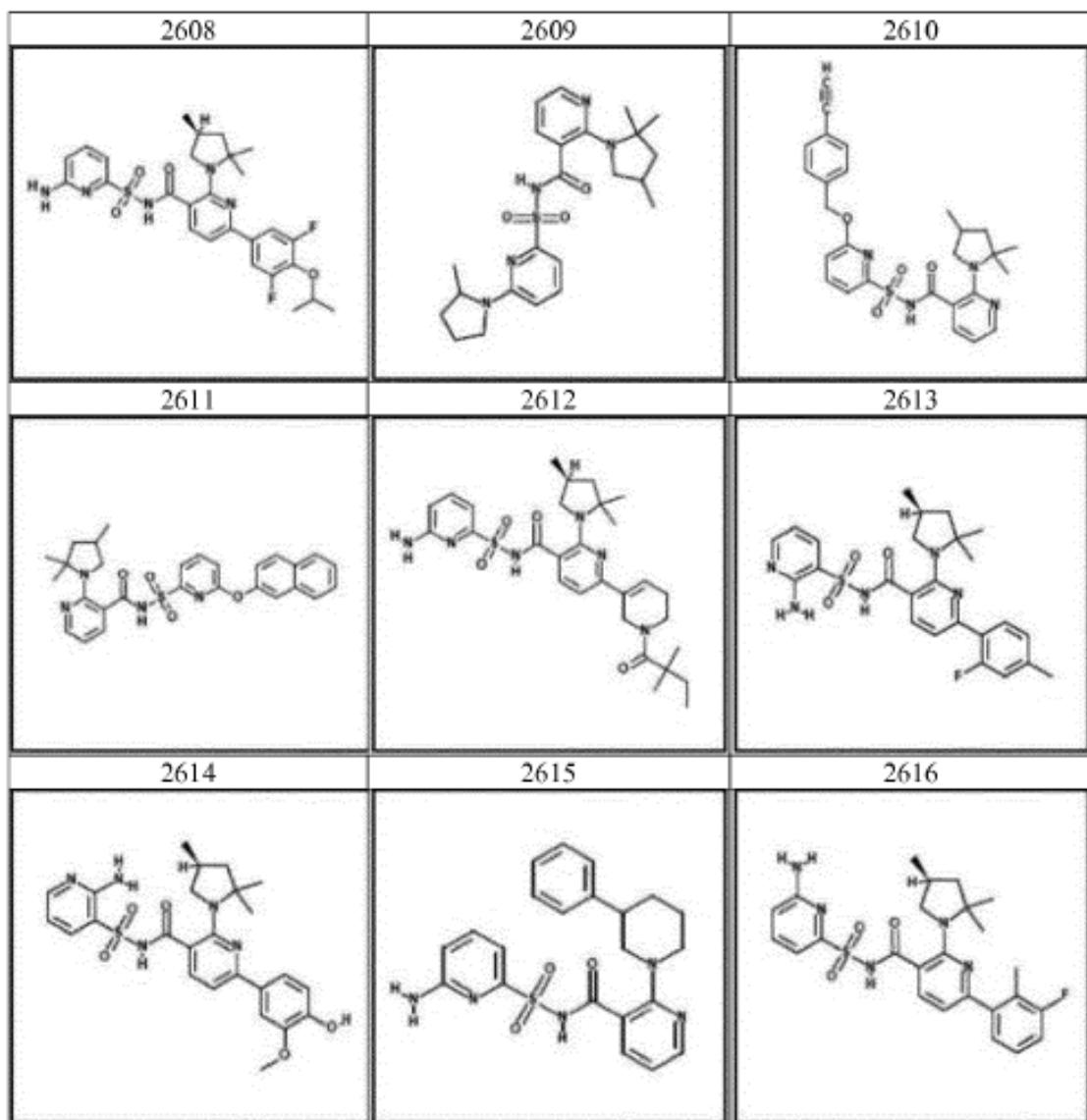


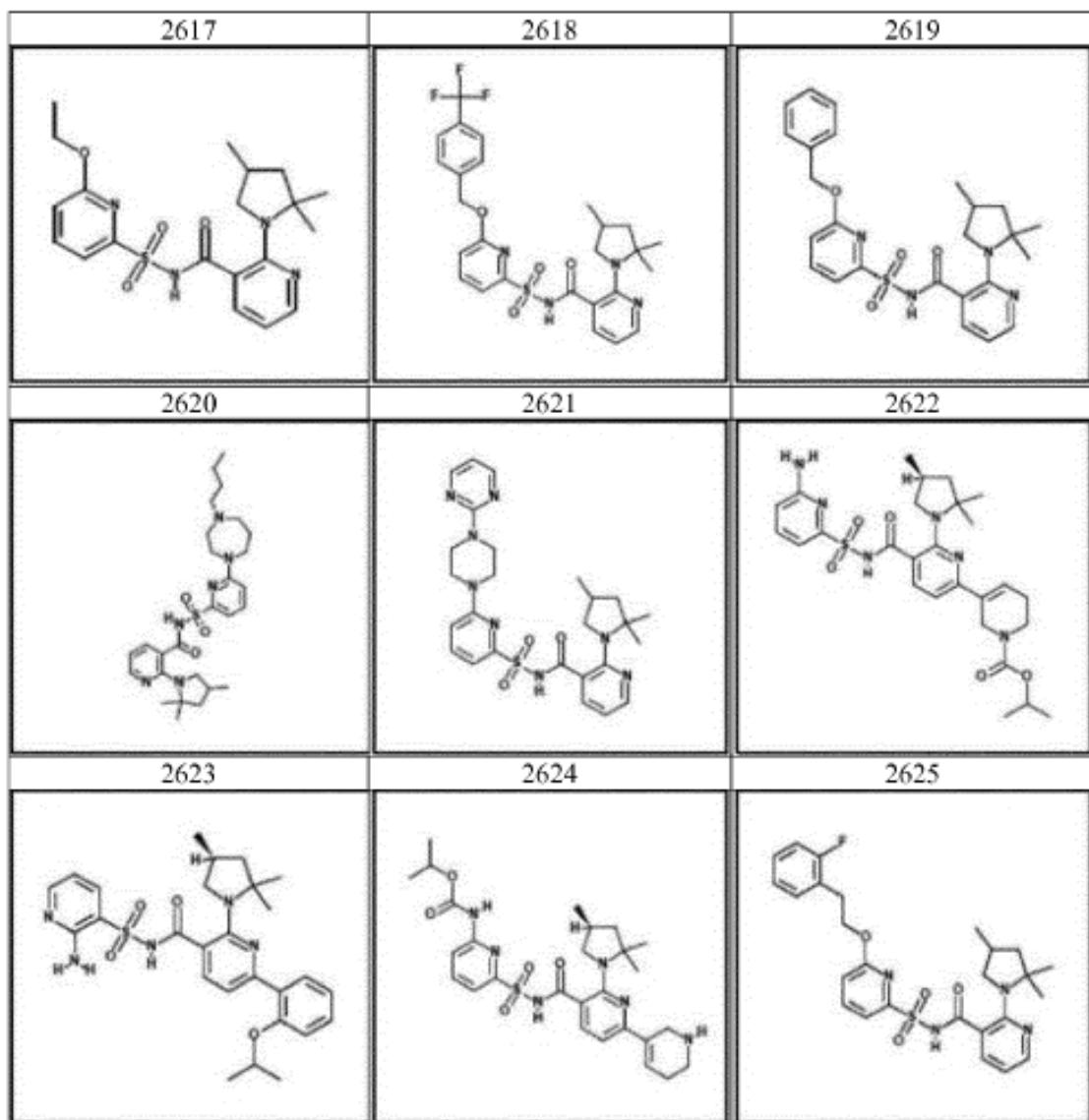


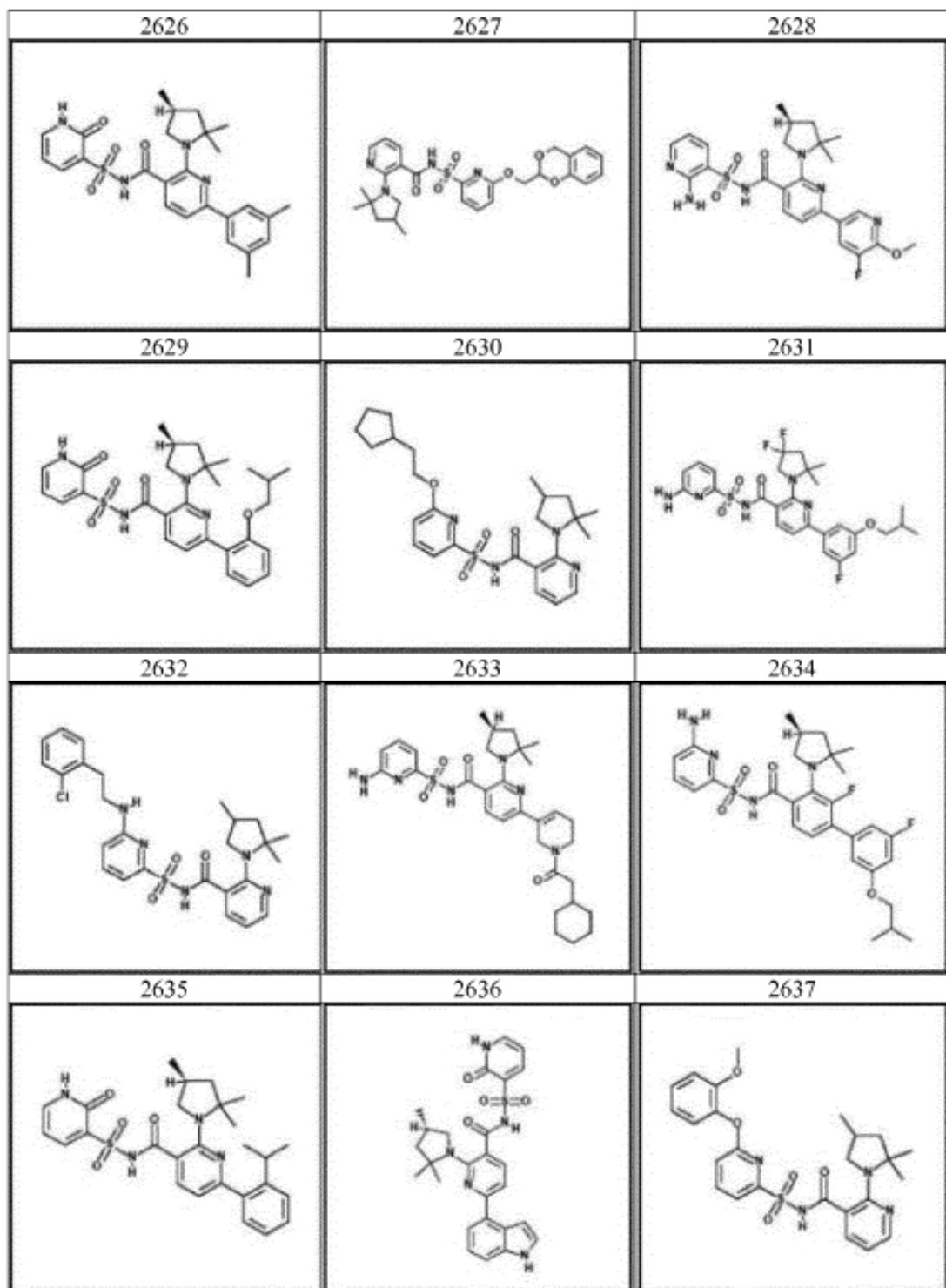


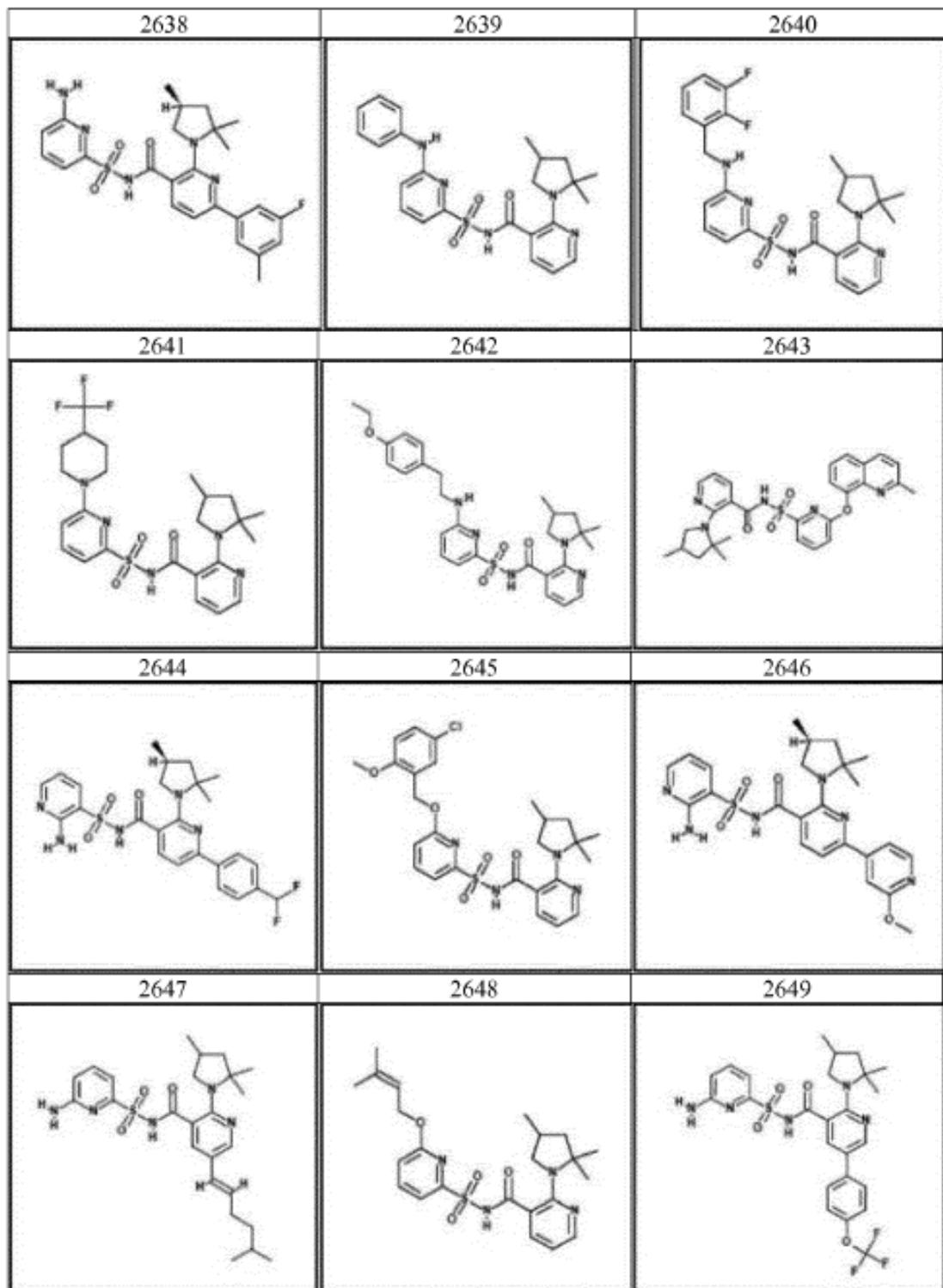


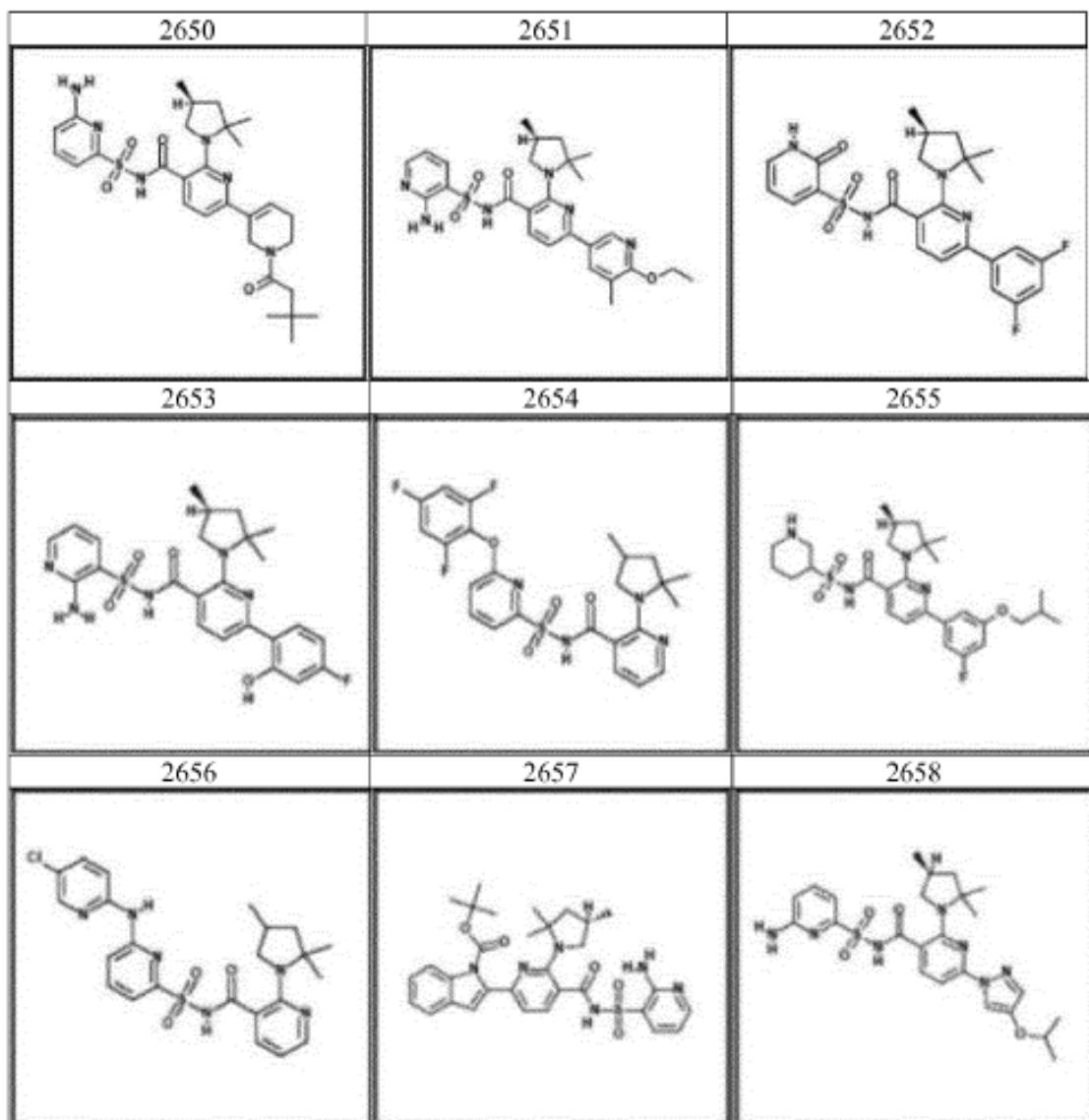


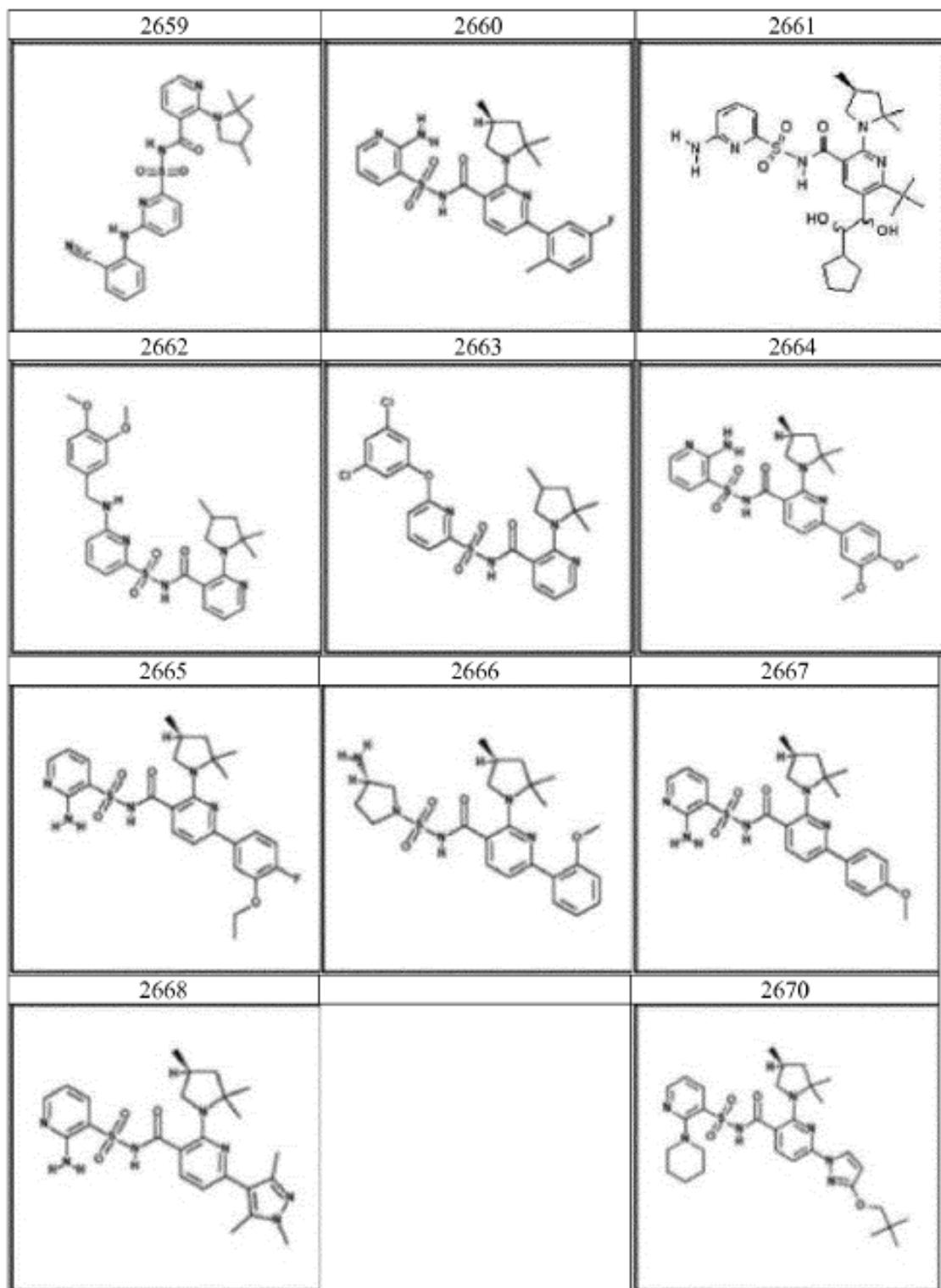




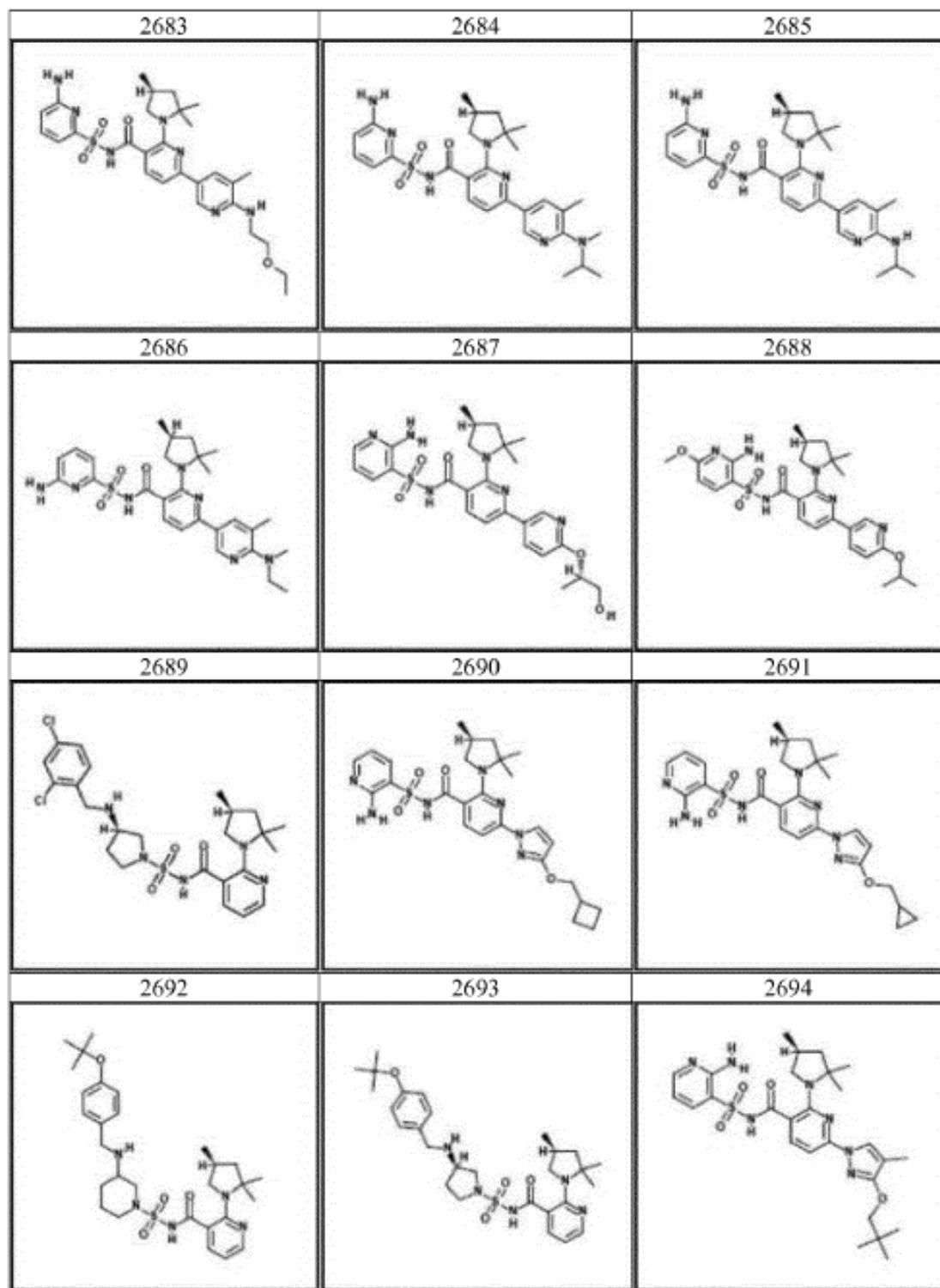


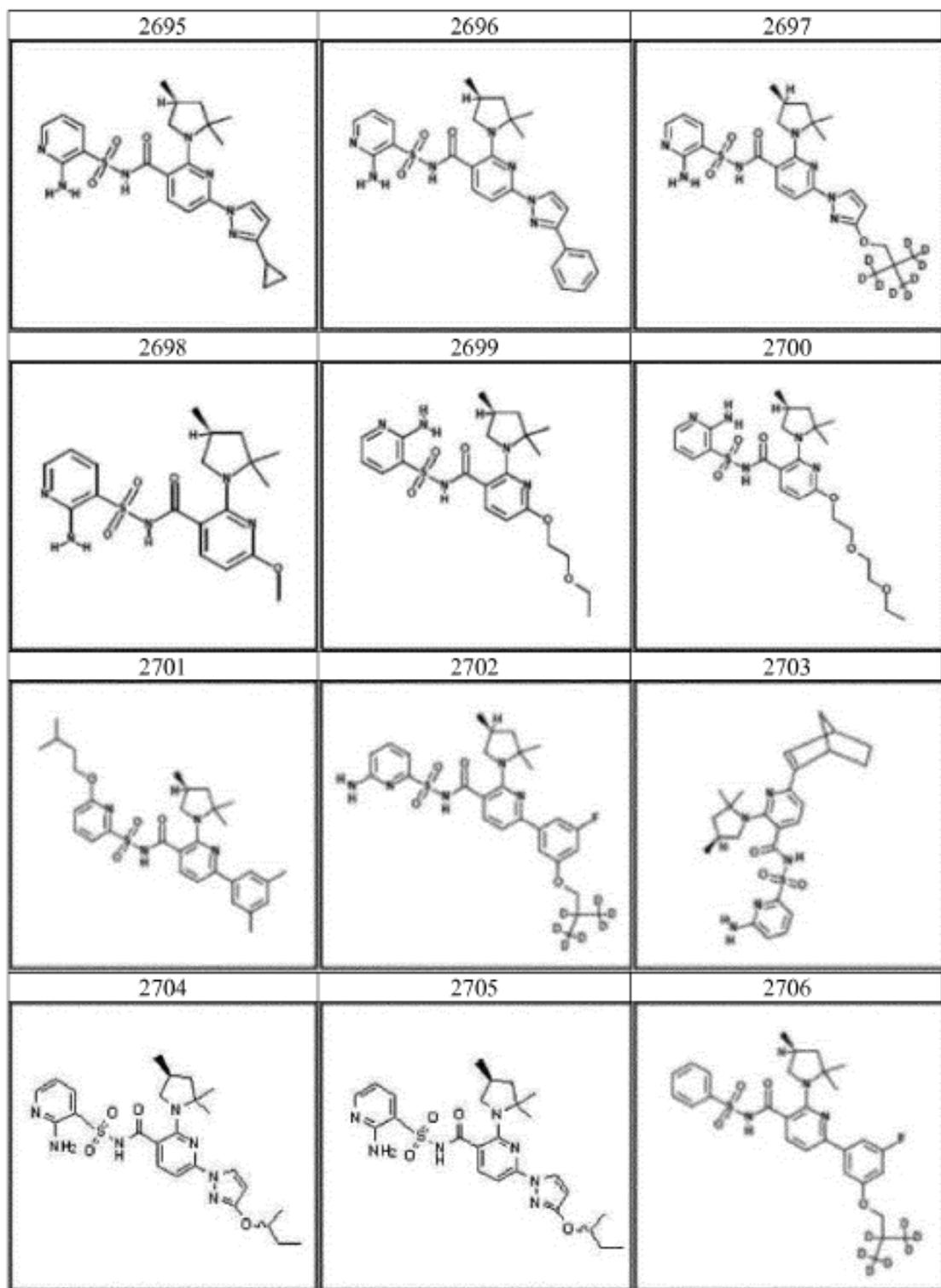


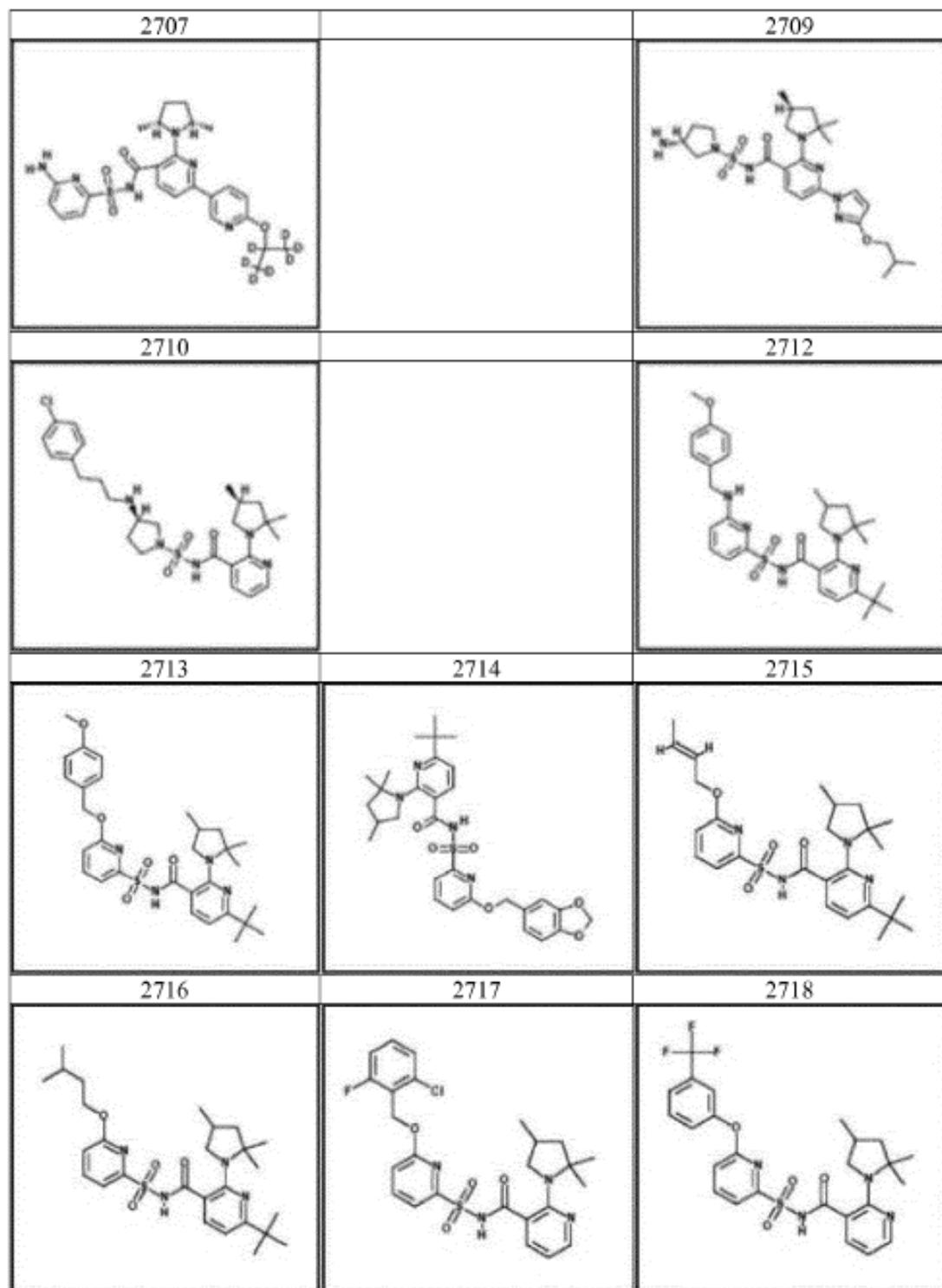


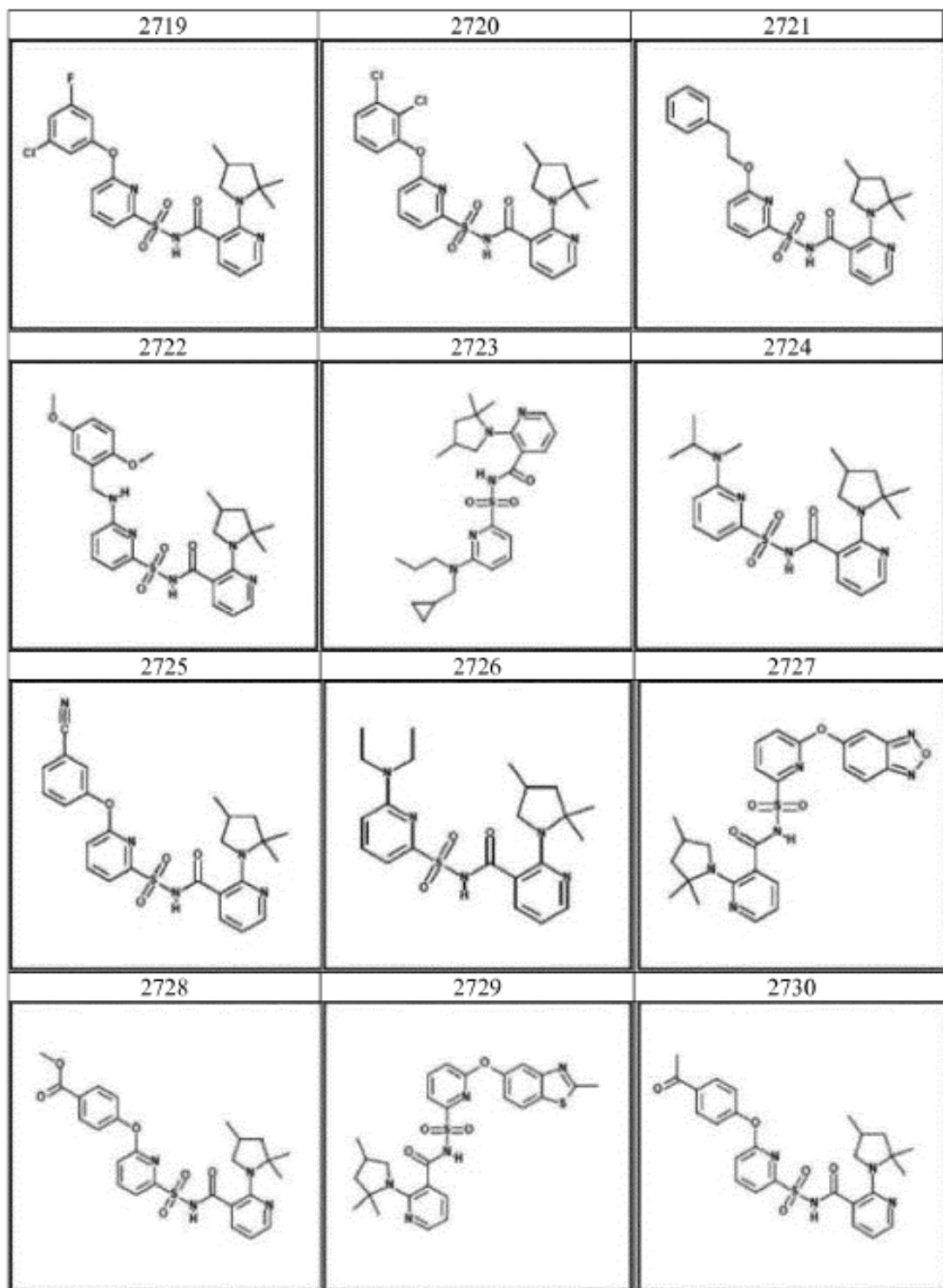


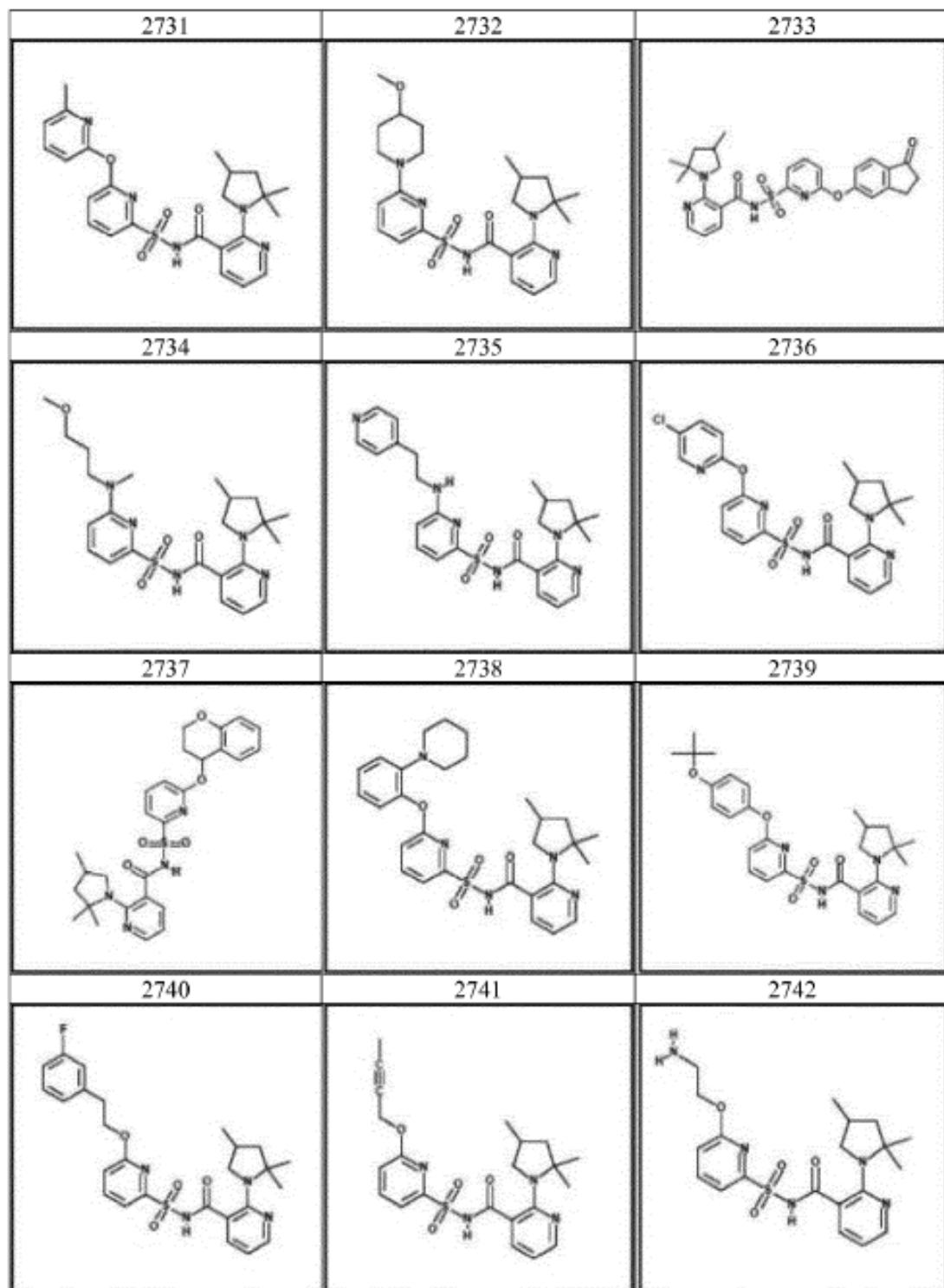
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2677	2678	2679
2680	2681	2682

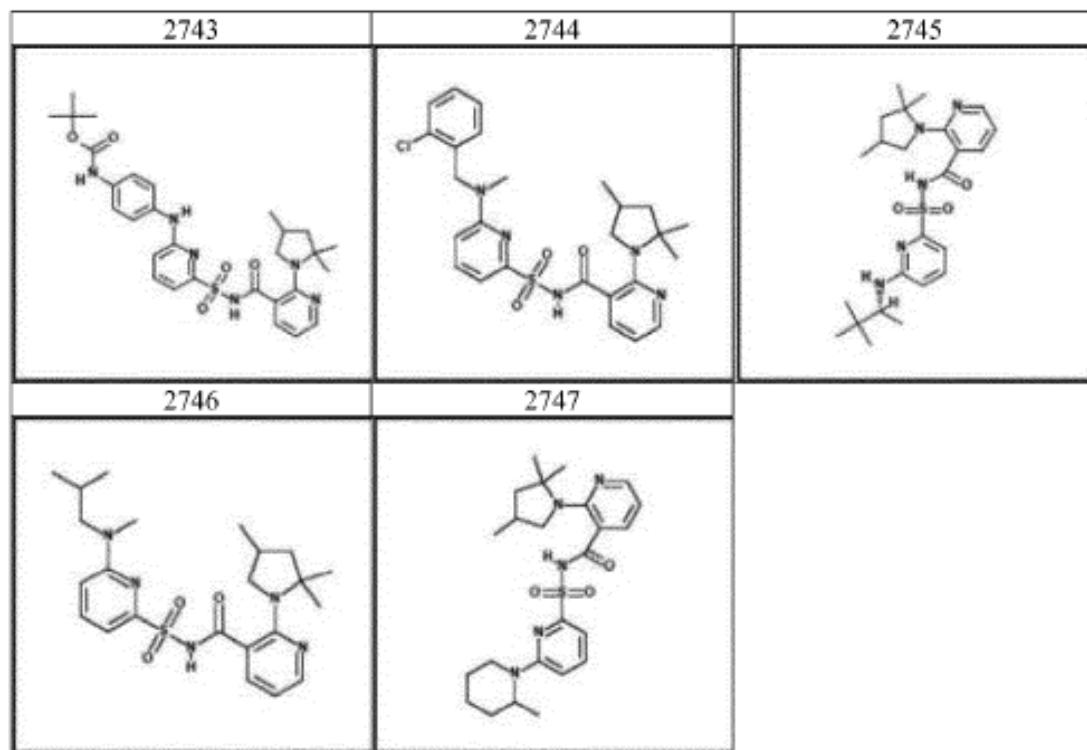




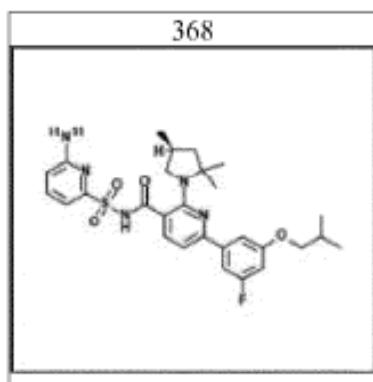






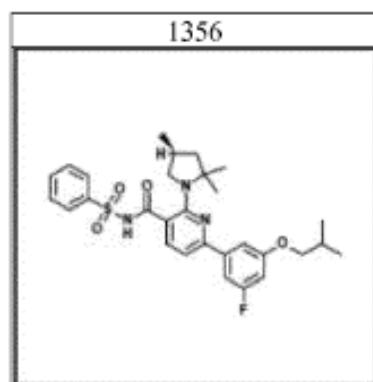


17. Forbindelse ifølge krav 16, eller et farmasøytisk akseptabelt salt derav, hvor forbindelsen er:



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18. Forbindelse ifølge krav 16, eller et farmasøytisk akseptabelt salt derav, hvor forbindelsen er:

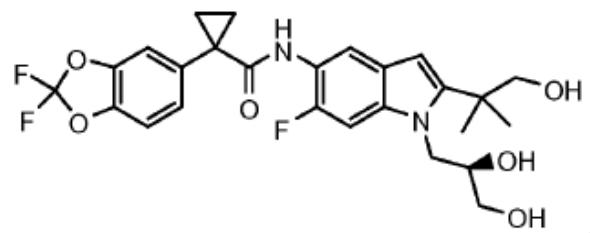


19. Farmasøytisk sammensetning omfattende forbindelse eller salt ifølge et hvilket som helst av kravene 1 til 18 og en farmasøytisk akseptabel bærer.

20. Farmasøytisk sammensetning ifølge krav 19, videre omfattende et eller flere ytterligere terapeutiske midler.

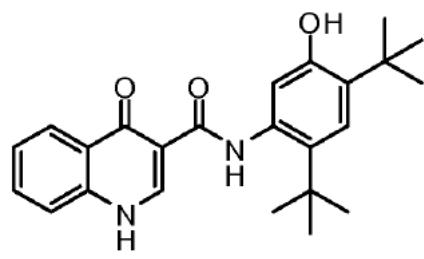
21. Farmasøytisk sammensetning ifølge krav 20, hvor det eller de ytterligere terapeutiske midlene omfatter en CFTR-modulator.

10 22. Farmasøytisk sammensetning ifølge et hvilket som helst av kravene 20 eller 21, hvor minst et ytterligere terapeutisk middel er



eller farmasøytisk akseptabelt salt derav.

15 23. Farmasøytisk sammensetning ifølge et hvilket som helst av kravene 20 til 22, hvor minst et ytterligere terapeutisk middel er



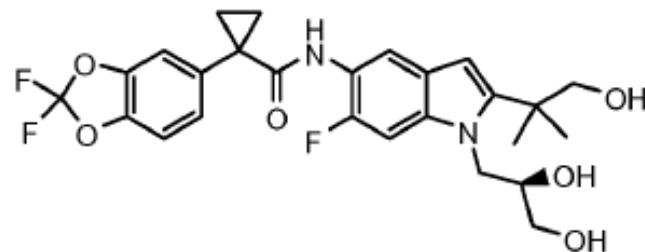
eller farmasøytisk akseptabelt salt derav.

20 24. Forbindelse eller salt ifølge et hvilket som helst av kravene 1 til 18, eller den farmasøytiske sammensetningen ifølge et hvilket som helst av kravene 19 til 23, for anvendelse i en fremgangsmåte for å behandle cystisk fibrose hos en pasient.

25. Forbindelse, salt eller sammensetning for anvendelse ifølge krav 24, hvor fremgangsmåten videre omfatter administrasjon til pasienten av et eller flere ytterligere terapeutiske midler før, samtidig med eller etter forbindelsen ifølge et hvilket som helst av kravene 1 til 18 eller den farmasøytiske sammensetningen ifølge et hvilket som helst av kravene 19 til 23.

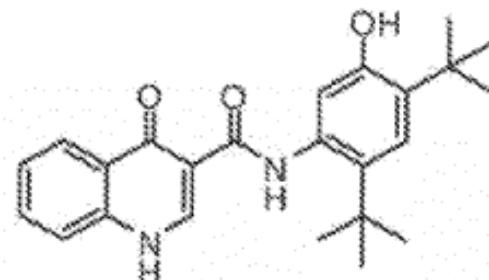
26. Forbindelse, salt eller sammensetning for anvendelse ifølge krav 25, hvor det ene eller flere ytterligere terapeutiske midler omfatter en CFTR-modulator.

27. Forbindelse, salt eller sammensetning for anvendelse ifølge et hvilket som helst av kravene 25 eller 26, hvor minst et ytterligere terapeutisk middel er



eller farmasøytisk akseptabelt salt derav.

28. Forbindelse, salt eller sammensetning for anvendelse ifølge et hvilket som helst av kravene 25 eller 26, hvor minst et ytterligere terapeutisk middel er:



29. Forbindelse, salt eller sammensetning for anvendelse ifølge et hvilket som helst av kravene 24 til 28, hvor pasienten er homozygot i F508del-mutasjonen; eller hvor pasienten er heterozygot i F508del-mutasjonen.