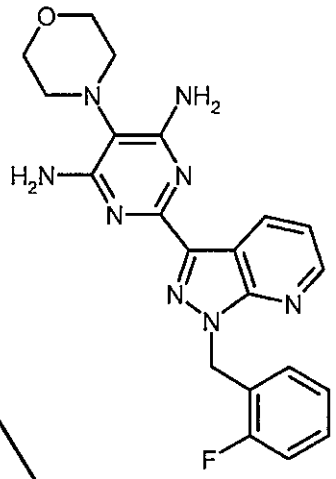
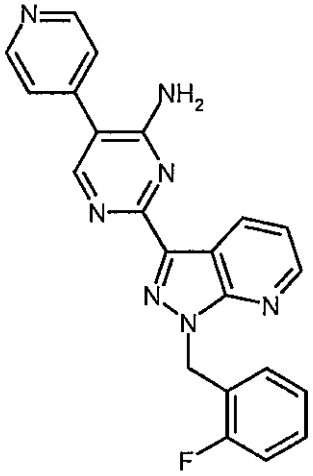
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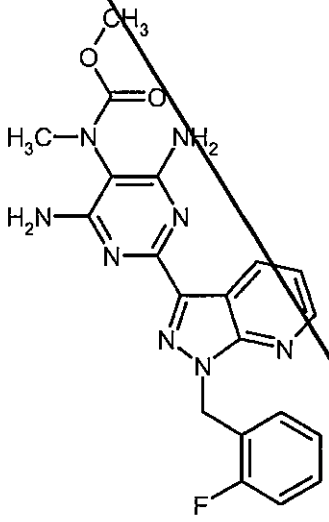
1. Compounds according to the formulae (1) to (27)



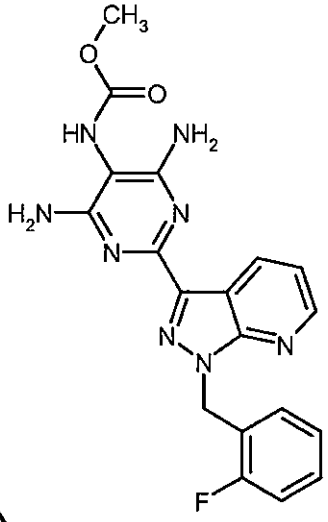
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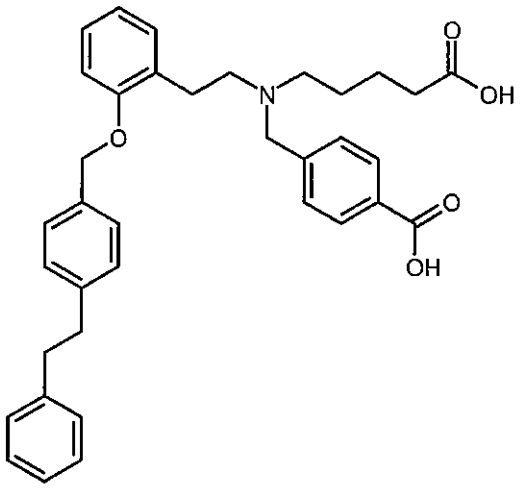
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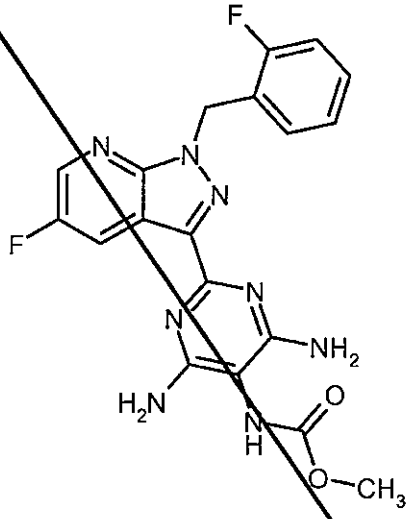
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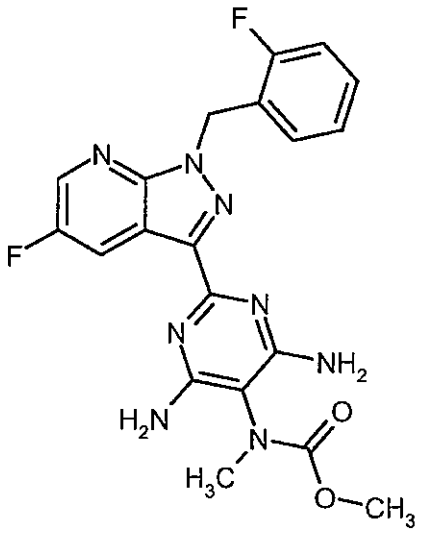
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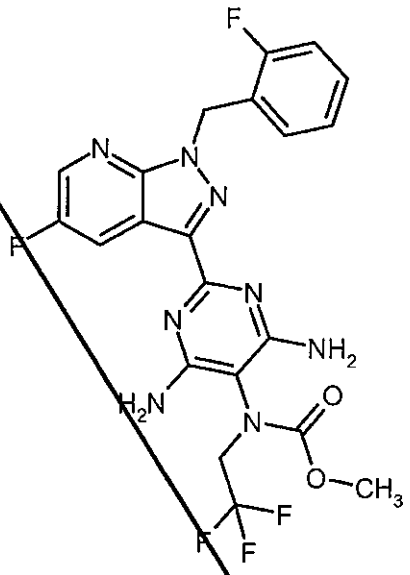
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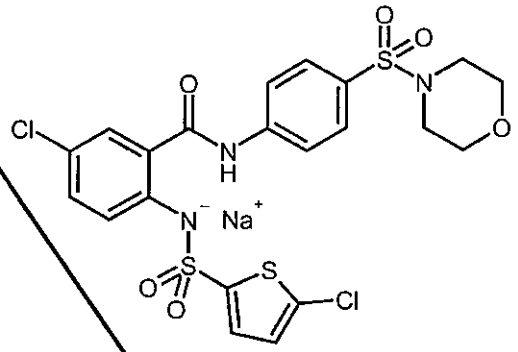
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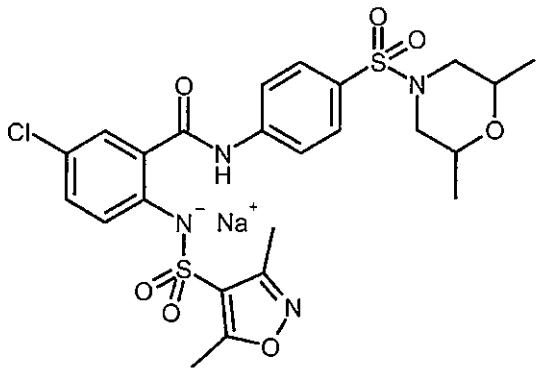
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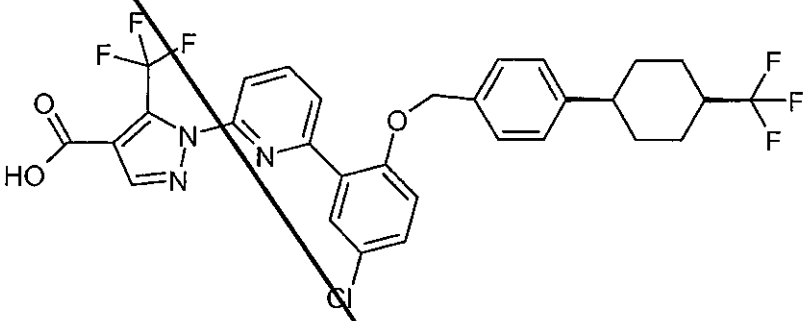
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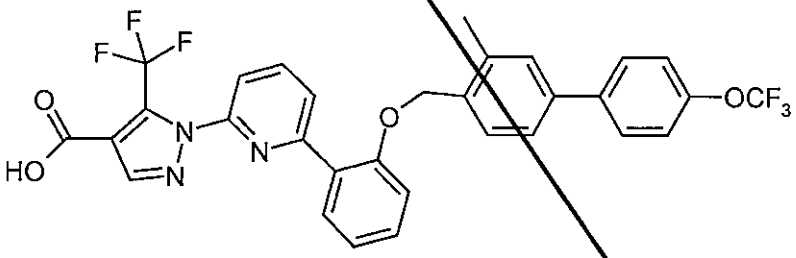
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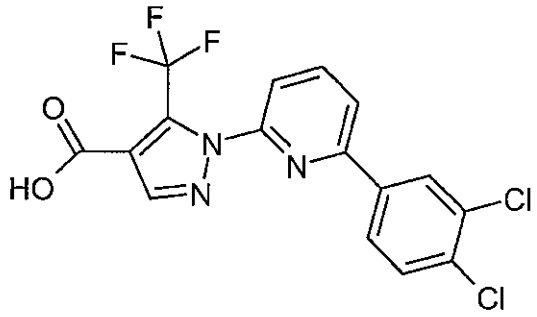
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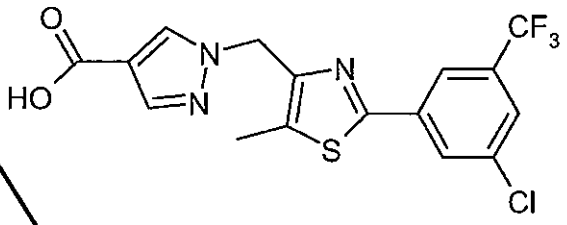
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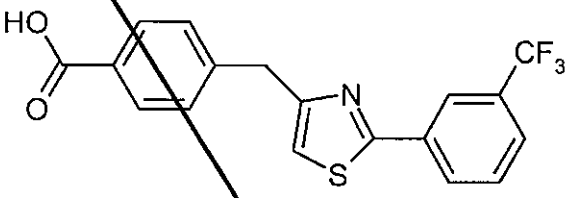
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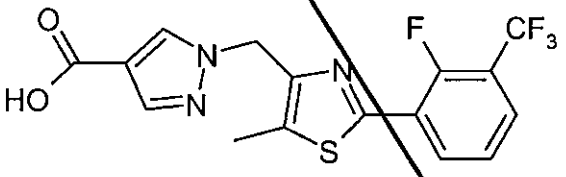
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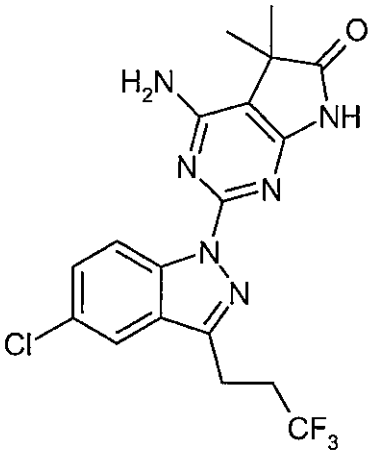
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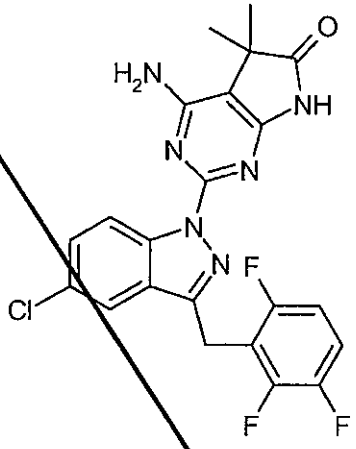
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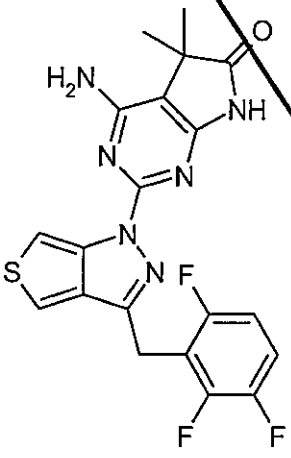
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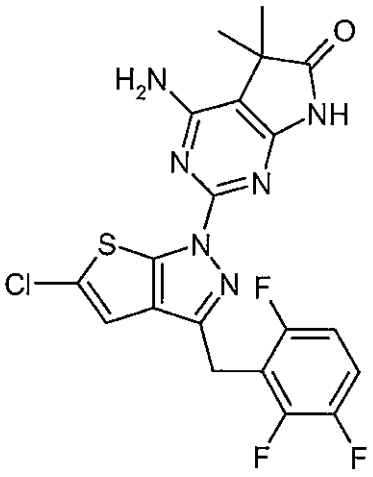
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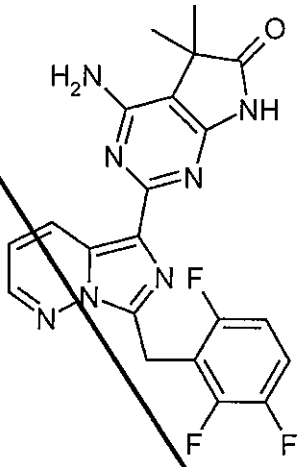
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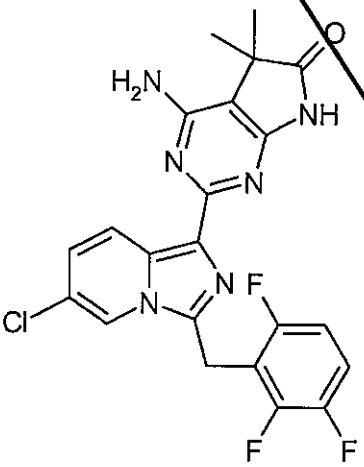
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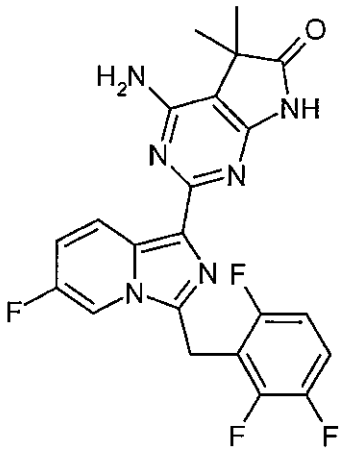
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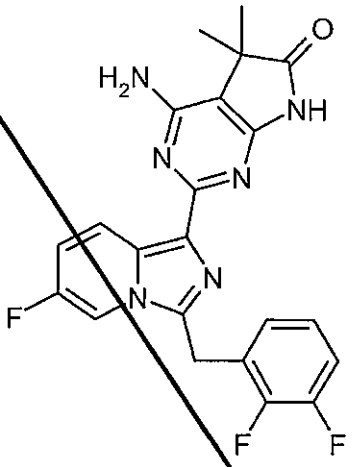
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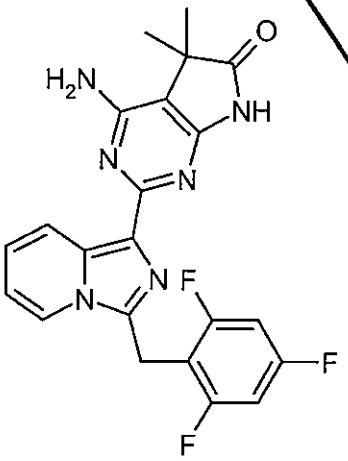
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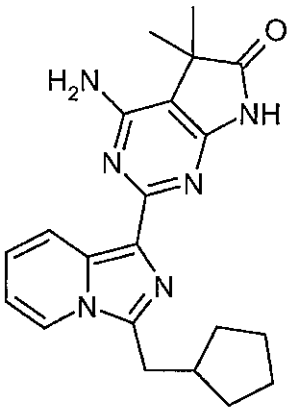
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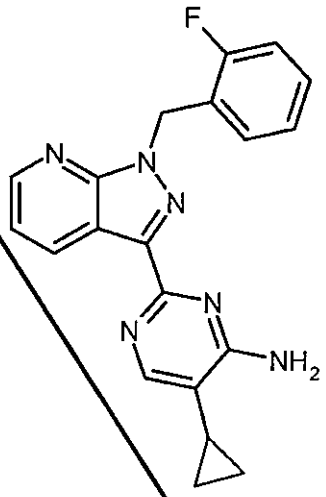
(24)



(25)



(26)



(27)

for use in the prevention and/or treatment of Systemic Sclerosis (SSc).

2. Compounds according to formulae and (1), (2), (3), (4), (5), (6), (7) and (27) for use in the prevention and/or treatment of Systemic Sclerosis (SSc).

3. At least one compound according to claim 1 and 2 in combination with at least one PDE5 Inhibitor selected from the group of:

Tadalafil ((6R,12aR) -2,3,6,7,12,12a – Hexahydro – 2 – methyl – 6 - (3,4-methylene - dioxyphenyl) pyrazino(1',2':1,6) pyrido(3,4-b)indole-1,4-dione), *Vardenafil* (2-(2-Ethoxy-5-(4-ethylpiperazin-1-yl-1-sulfonyl)phenyl)-5-methyl-7-propyl-3H-imidazo (5,1-f) (1,2,4)triazin-4-one), *Sildenafil* (3-[2-ethoxy-5-(4-methylpiperazin-1-yl)sulfonyl-phenyl]-7- methy l- 9- propy l-2,4,7,8- tetrazabicyclo [4.3.0]nona -3,8,10-trien-5-one), *Udenafil* 5-[2-propyloxy-5-(1-methyl-2-pyrrolidinylethylamidosulfonyl)phenyl]-methyl-3-propyl-1,6-

dihydro-7H-pyrazolo(4,3-d)pyrimidine-7-one, *Dasantafil* 7-(3-Bromo-4-methoxybenzyl)-1-ethyl-8-[[(1,2)-2-hydroxycyclopentyl]amino]-3-(2-hydroxyethyl)-3,7-dihydro-1-purine-2,6-dione, *Avanafil* 4-{[(3-chloro-4-methoxyphenyl)methyl]amino}-2-[(2S)-2-(hydroxymethyl)pyrrolidin-1-yl]-N-(pyrimidin-2-ylmethyl)pyrimidine-5-carboxamide, Mirodenafil, Lodenafil, UK 369.003, UK 371.800, *SLx 2101* of Surface Logix, *LAS 34179*Triazolo[1,2-]xanthine,6-methyl-4-propyl-2-[2-propoxy-5-(4-methylpiperazino)sulfonyl]phenyl or salts, hydrates or hydrates of the salts

for use in the prevention and/or treatment of Systemic Sclerosis (SSc).

4. At least one compound according to claim 1 and 2 in combination with sildenafil or vardenafil for use in the prevention and/or treatment of Systemic Sclerosis (SSc).

5. Compounds according to claim 1 to 4 for use in the prevention and/or treatment of Systemic Sclerosis (SSc), diffuse Systemic Sclerosis (dSSc), limited Systemic Sclerosis (lSSc), overlap type of Systemic Sclerosis, undifferentiated type of Systemic Sclerosis, Systemic Sclerosis sine scleroderma, skin fibrosis, scleroderma, nephrogenic fibrosing dermopathy (NFD), nephrogenic systemic fibrosis (NSF), keloid formation.

6. Compounds according to claim 1 to 4 for use in the prevention and/or treatment of Systemic Sclerosis SSc concomitant fibrosis of internal organs, comprising the gut, the lung, the kidney and the blood vessels.

7. Compounds according to formulae 3, 4, 6, and/or 7 for use in the prevention and/or treatment of Systemic Sclerosis SSc.

8. At least one compound according to formulae 3, 4, 6, and/or 7 in combination with vardenafil or sildenafil for use in the prevention and/or treatment of Systemic Sclerosis SSc.

9. Use for the production of a medicament for prevention and/or treatment of Systemic Sclerosis (SSc) comprising an effective amount of a compound according to claim 1 to 4.

10. Use for the production of a medicament for prevention and/or treatment of Systemic Systemic Sclerosis (SSc), diffuse Systemic Sclerosis (dSSc), limited Systemic Sclerosis (lSSc), overlap type of Systemic Sclerosis, undifferentiated type of Systemic Sclerosis, Systemic Sclerosis sine scleroderma, skin fibrosis, scleroderma, nephrogenic fibrosing dermopathy (NFD), keloid formation comprising an effective amount of a compound according to 1 to 4.

11. Use for the production of a medicament for prevention and/or treatment of Systemic Sclerosis SSc concomitant fibrosis of internal organs, comprising the gut, the lung, the kidney and the blood vessels comprising an effective amount of a compound according to 1 to 4.

12. Pharmaceutical formulation comprising at least one compound or one combination according to claims 1 to 4 for the use in the prevention and/or treatment of Systemic Sclerosis (SSc).

13. Pharmaceutical formulation comprising at least one compound or one combination according to claims 1 to 4 for the use in the prevention and/or treatment of Systemic Sclerosis (SSc), diffuse Systemic Sclerosis (dSSc), limited Systemic Sclerosis (lSSc), overlap type of Systemic Sclerosis, undifferentiated type of Systemic Sclerosis, Systemic Sclerosis sine scleroderma, skin fibrosis, scleroderma, nephrogenic fibrosing dermopathy (NFD), keloid formation.

14. Pharmaceutical formulation comprising at least one compound or one combination according to claims 1 to 4 for the use in the prevention and/or treatment of Systemic Sclerosis SSc concomitant fibrosis of internal organs, comprising the gut, the lung, the kidney and the blood vessels.

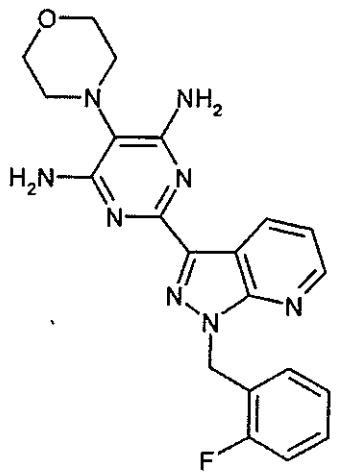
15. Kit comprising at least one sGC stimulator and/or activator according to claims 1 to 2 or a combination according to claims 3 to 4 for the use in the prevention and/or treatment of Systemic Sclerosis (SSc).

16. Kit according to claim 13 for the use in the prevention and/or treatment of Systemic Sclerosis (SSc), diffuse Systemic Sclerosis (dSSc), limited Systemic Sclerosis (lSSc), overlap type of Systemic Sclerosis, undifferentiated type of Systemic Sclerosis, Systemic Sclerosis sine scleroderma, skin fibrosis, scleroderma, nephrogenic fibrosing dermopathy (NFD), keloid formation.

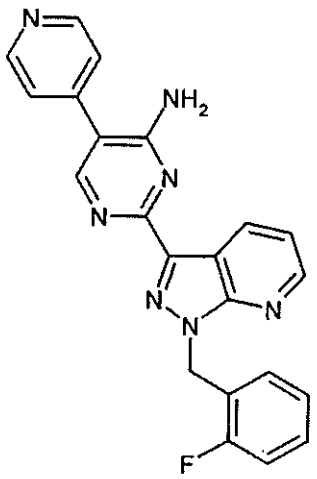
17. Kit according to claims 13 and 14 for the use in the prevention and/or treatment of Systemic Sclerosis SSc concomitant fibrosis of internal organs, comprising the gut, the lung, the kidney and the blood vessels.

Claims

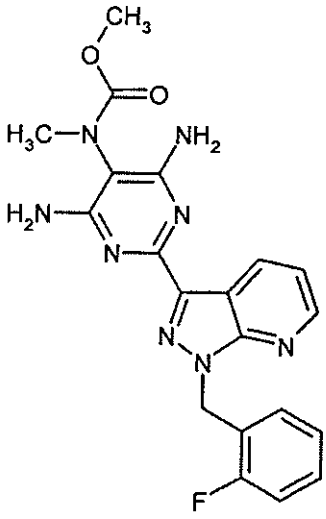
1. Compounds according to the formulae (1) to (27)



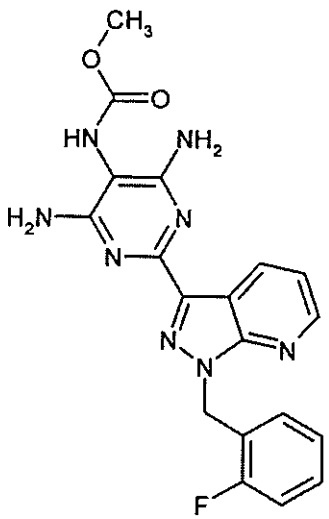
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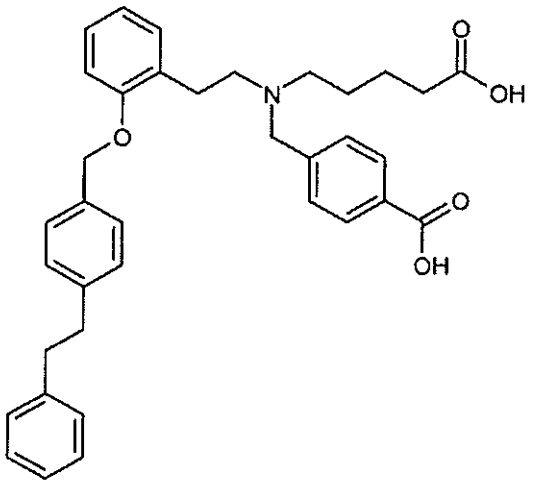
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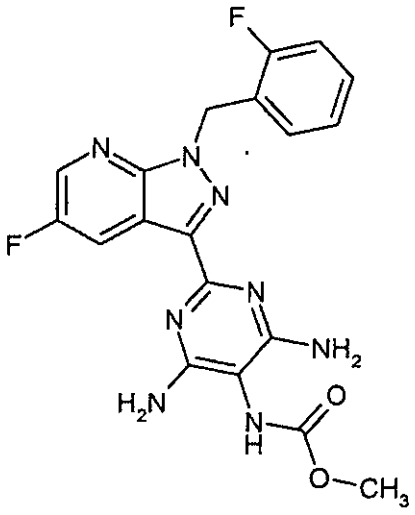
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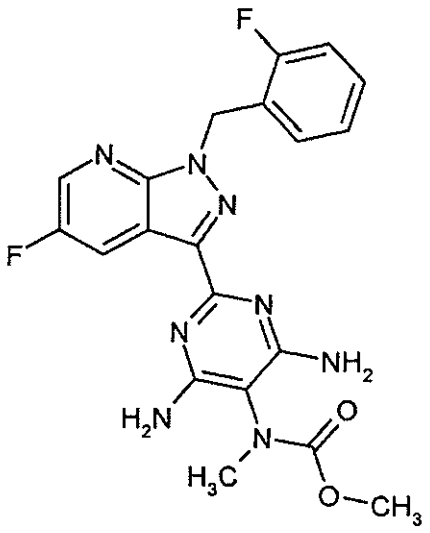
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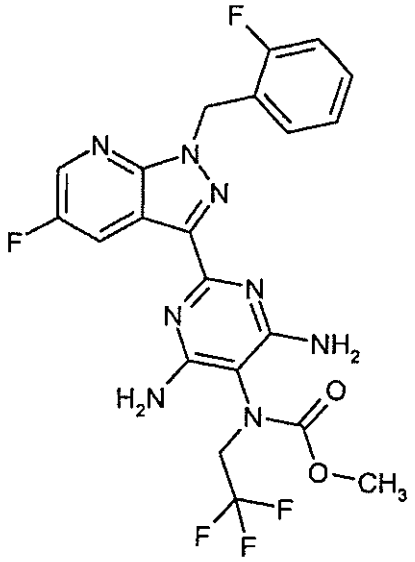
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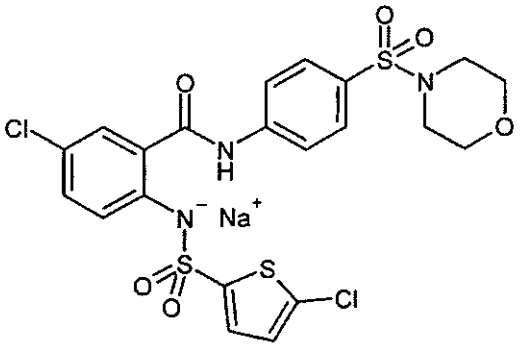
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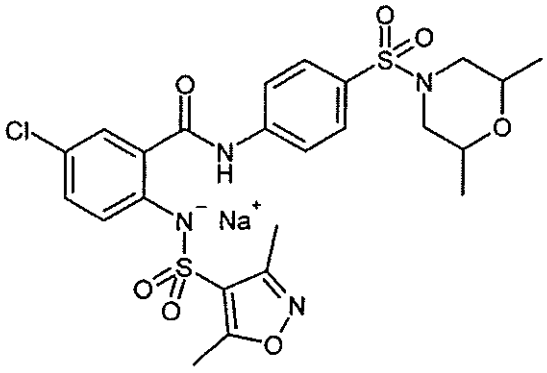
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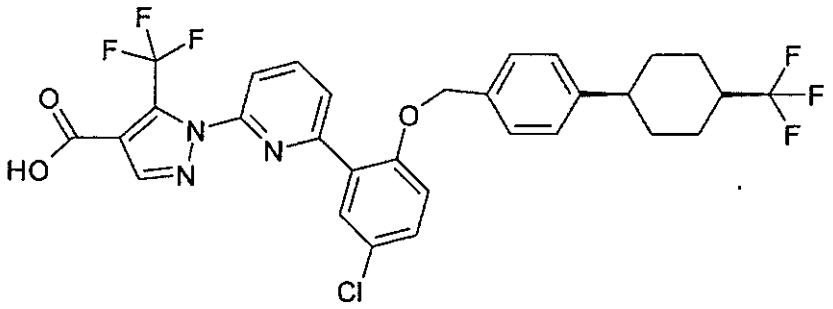
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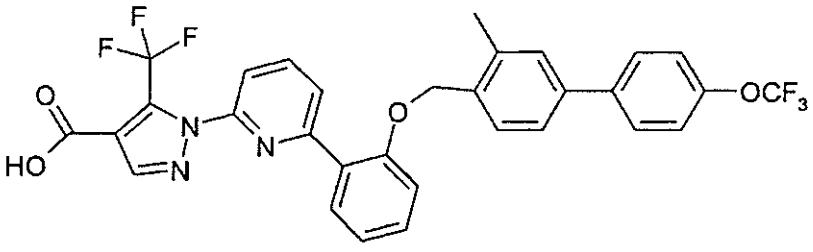
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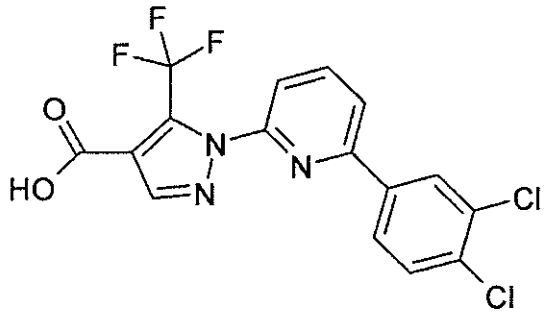
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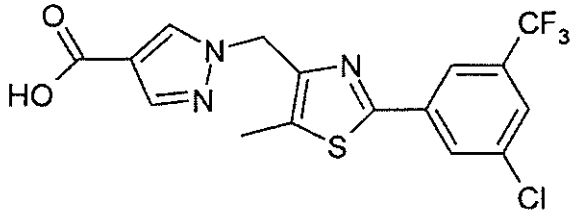
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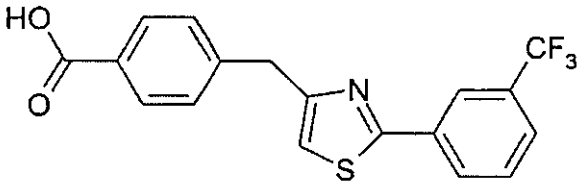
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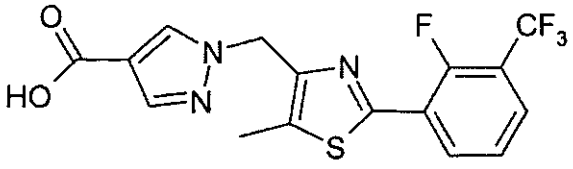
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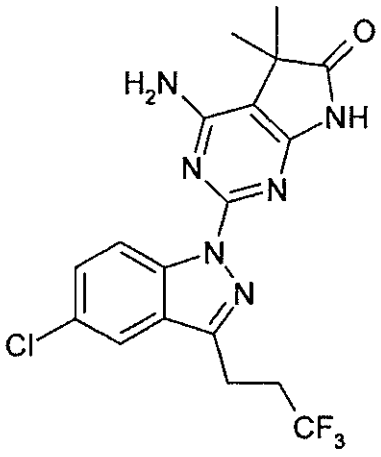
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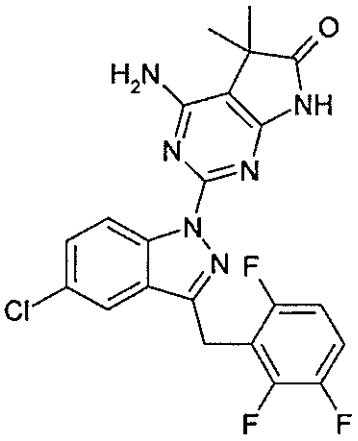
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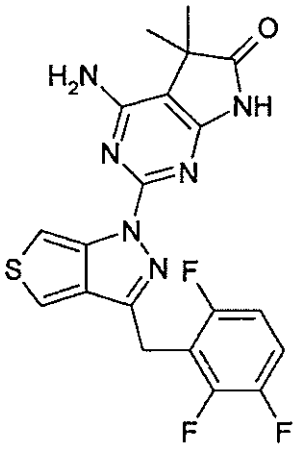
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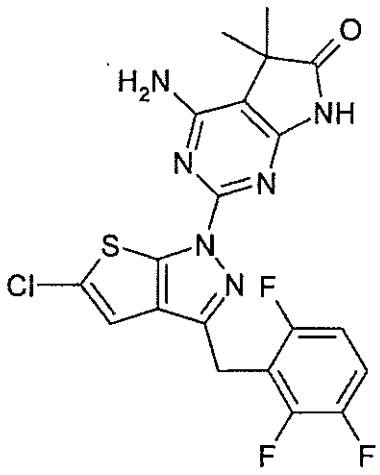
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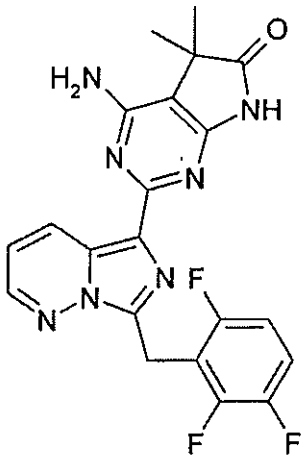
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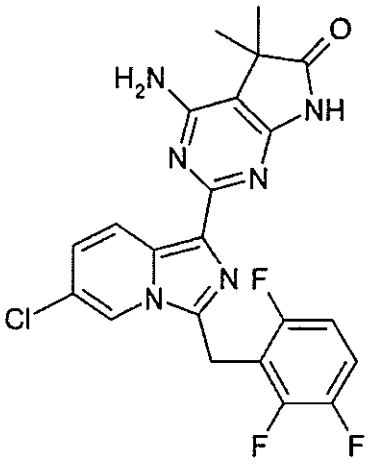
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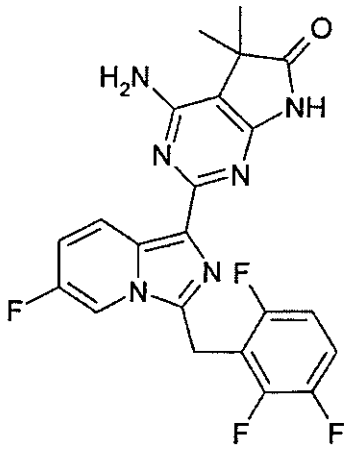
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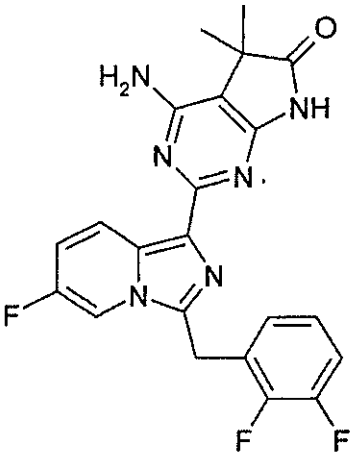
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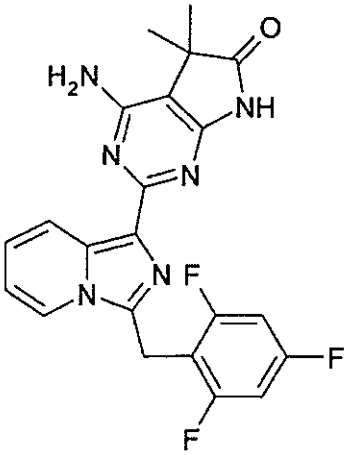
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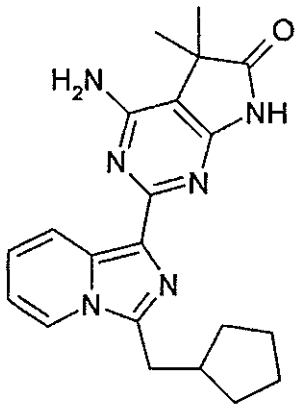
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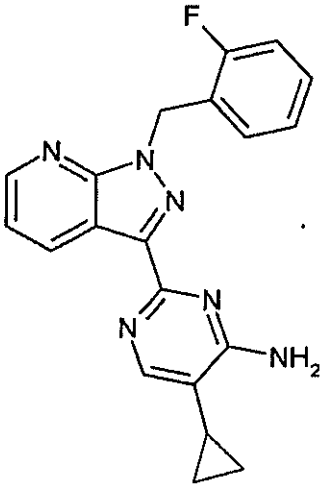
(24)



(25)



(26)



(27)

for use in the prevention and/or treatment of Systemic Sclerosis (SSc).

2. Compounds according to formulae and (1), (2), (3), (4), (5), (6), (7) and (27) for use in the prevention and/or treatment of Systemic Sclerosis (SSc).

3. Compound according to formula (3) for use in the prevention and/or treatment of Systemic Sclerosis.

4. At least one compound according to claim 1 to 3 in combination with at least one PDE5 Inhibitor selected from the group of:

   Tadalafil ((6R,12aR) -2,3,6,7,12,12a – Hexahydro – 2 – methyl – 6 - (3,4-methylene - dioxyphenyl) pyrazino(1',2':1,6) pyrido(3,4-b)indole-1,4-dione), *Vardenafil* (2-(2-Ethoxy-

5-(4-ethylpiperazin-1-yl-1-sulfonyl)phenyl)-5-methyl-7-propyl-3H-imidazo (5,1-f) (1,2,4)triazin-4-one), *Sildenafil* (3-[2-ethoxy-5-(4-methylpiperazin-1-yl)sulfonyl-phenyl]-7- methy l- 9- propy l-2,4,7,8- tetrazabicyclo [4.3.0]nona -3,8,10-trien-5-one), *Udenafil* 5-[2-propyloxy-5-(1-methyl-2-pyrrolidinylethylamidosulfonyl)phenyl]-methyl-3-propyl-1,6-dihydro-7H-pyrazolo(4,3-d)pyrimidine-7-one, *Dasantafil* 7-(3-Bromo-4-methoxybenzyl)-1-ethyl-8-[[(1,2)-2-hydroxycyclopentyl]amino]-3-(2-hydroxyethyl)-3,7-dihydro-1-purine-2,6-dione, *Avanafil* 4-{[(3-chloro-4-methoxyphenyl)methyl]amino}-2-[(2S)-2-(hydroxymethyl)pyrrolidin-1-yl]-N-(pyrimidin-2-ylmethyl)pyrimidine-5-carboxamide, Mirodenafil, Lodenafil, UK 369.003, UK 371.800, *SLx 2101* of Surface Logix, *LAS 34179*Triazolo[1,2-]xanthine,6-methyl-4-propyl-2-[2-propoxy-5-(4-methylpiperazino)sulfonyl]phenyl or salts, hydrates or hydrates of the salts

for use in the prevention and/or treatment of Systemic Sclerosis (SSc).

5. At least one compound according to claim 1 to 3 in combination with sildenafil or vardenafil for use in the prevention and/or treatment of Systemic Sclerosis (SSc).
6. Compounds according to claim 1 to 5 for use in the prevention and/or treatment of Systemic Sclerosis (SSc), diffuse Systemic Sclerosis (dSSc), limited Systemic Sclerosis (lSSc), overlap type of Systemic Sclerosis, undifferentiated type of Systemic Sclerosis, Systemic Sclerosis sine scleroderma, skin fibrosis, scleroderma, nephrogenic fibrosing dermopathy (NFD), nephrogenic systemic fibrosis (NSF), keloid formation.
7. Compounds according to claim 1 to 5 for use in the prevention and/or treatment of Systemic Sclerosis SSc concomitant fibrosis of internal organs, comprising the guts, the lungs, the kidneys

and the blood vessels.

8. Compounds according to formulae (3), (4), (6), and/or (7) for use in the prevention and/or treatment of Systemic Sclerosis SSc.
9. At least one compound according to formulae (3), (4), (6) and/or (7) in combination with

vardenafil or sildenafil for use in the prevention and/or treatment of Systemic Sclerosis SSc.

10. Use of a compound according to claims 1-9 for

the production of a medicament for prevention and/or treatment of Systemic Sclerosis (SSc) comprising an effective amount of a compound according to claims 1 to 9.

11. Use of a compound according to claims 1-9 for

the production of a medicament for prevention and/or treatment of Systemic Systemic Sclerosis (SSc), diffuse Systemic Sclerosis (dSSc), limited Systemic Sclerosis (lSSc), overlap type of Systemic Sclerosis, undifferentiated type of Systemic Sclerosis,

Systemic Sclerosis sine scleroderma, skin fibrosis, scleroderma, nephrogenic fibrosing dermopathy (NFD), keloid formation comprising an effective amount of a compound according to claims 1 to 9.

12. Use of a compound according to claims 1-9 for the production of a medicament for prevention and/or treatment of Systemic Sclerosis SSc concomitant fibrosis of internal organs, comprising the guts, the lungs, the kidneys and the blood vessels comprising an effective amount of a compound according to claims 1 to 9.

13. Pharmaceutical formulation comprising at least one compound or one combination according to claims 1 to 9 for the use in the prevention and/or treatment of Systemic Sclerosis (SSc).

14. Pharmaceutical formulation comprising at least one compound or one combination according to claims 1 to 9 for the use in the prevention and/or treatment of Systemic Sclerosis (SSc), diffuse Systemic Sclerosis (dSSc), limited Systemic Sclerosis (lSSc), overlap type of Systemic Sclerosis, undifferentiated type of Systemic Sclerosis, Systemic Sclerosis sine scleroderma, skin fibrosis, scleroderma, nephrogenic fibrosing dermopathy (NFD), keloid formation.

15. Pharmaceutical formulation comprising at least one compound or one combination according to claims 1 to 9 for the use in the prevention and/or treatment of Systemic Sclerosis SSc concomitant fibrosis of internal organs, comprising the guts, the lungs, the kidneys and the blood vessels.

16. Kit comprising at least one sGC stimulator and/or activator according to claims 1 to 3 or a combination according to claims 4 to 5 for the use in the prevention and/or treatment of Systemic Sclerosis (SSc).

17. Kit according to claim 16 for the use in the prevention and/or treatment of Systemic Sclerosis (SSc), diffuse Systemic Sclerosis (dSSc), limited Systemic Sclerosis (lSSc), overlap type of Systemic Sclerosis, undifferentiated type of Systemic Sclerosis, Systemic Sclerosis sine scleroderma, skin fibrosis, scleroderma, nephrogenic fibrosing dermopathy (NFD), keloid formation.

18. Kit according to claims 16 and 17 for the use in the prevention and/or treatment of Systemic Sclerosis SSc concomitant fibrosis of internal organs, comprising the guts, the lungs, the kidneys and the blood vessels.