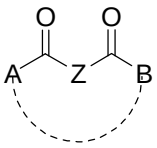
Claims

1. A compound according to the invention is given by Formula (I)

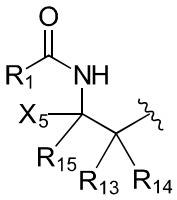


(I)

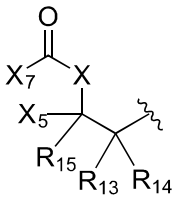
or a pharmaceutically acceptable salt thereof, wherein the dotted bond between A and B denotes an optional bond so as to form a ring closed structure, and wherein

Z is–CH_2-CH_2-,

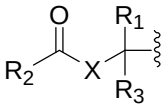
A is -SR, and R is

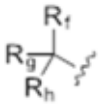


or

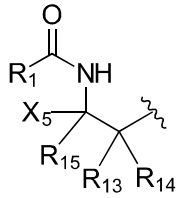


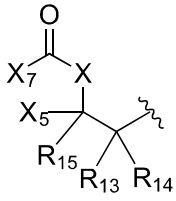
B is selected from -O-R', -SR''' or -OH; and R' is selected from the formulae:



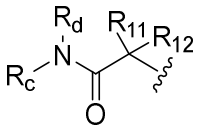


and R''' are independently different or identical and is selected from the formulae below:





or



R_1 is selected from H, Me, Et, propyl, i-propyl, butyl, iso-butyl, t-butyl, O-acyl, O-alkyl, N-acyl, N-alkyl, Xacyl, CH_2Xalkyl, CH_2X-acyl, F, CH_2COOH, CH_2CO_2alkyl,

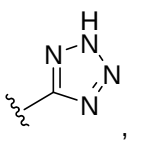
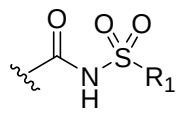
R_3 is selected from H, Me, Et, propyl, i-propyl, t-butyl, butyl, iso-butyl, O-acyl, O-alkyl, N-acyl, N-alkyl, Xacyl, CH_2Xalkyl, CH_2X-acyl, F, CH_2COOH, CH_2CO_2alkyl,

X is selected from O, NH, S,

R_2 is selected from Me, Et, propyl, i-propyl, butyl, iso-butyl, t-butyl, $C(O)CH_3$, $C(O)CH_2C(O)CH_3$, $C(O)CH_2CH(OH)CH_3$,

R_6 is selected from H, Me, Et, propyl, i-propyl, butyl, iso-butyl, t-butyl, acetyl, acyl, propionyl or benzoyl,

X_5 is selected from -H**,** Me, Et, propyl, i-propyl, butyl, iso-butyl, t-butyl, -COOH, $-C(=O)XR_6$, $CONR_1R_3$ or is formula


,


X_7 is selected from R_1, $-NR_1R_3$,

R_9 is selected from H, Me, Et or $O_2CCH_2CH_2COXR_8$

R_8 is selected from H, alkyl, Me, Et, propyl, i-propyl, butyl, iso-butyl, t-butyl, acetyl, acyl, propionyl, benzoyl, or formula (II),

R_{11} and R_{12} are independently different or identical and are selected from H, Me, Et, propyl, i-propyl, butyl, iso-butyl, t-butyl, acetyl, propionyl, benzoyl, $-CH_2Xalkyl$, $-CH_2Xacyl$, where X is O, NR_6 or S,

R_c and R_d is selected from $CH_2Xalkyl$, CH_2Xacyl, where X = O, NR_6 or S,

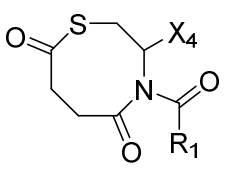
R_{13}, R_{14} and R_{15} are independently different or identical and are selected from H, Me, Et, propyl, i-propyl, butyl, iso-butyl, t-butyl, -COOH, O-acyl, O-alkyl, N-acyl, N-alkyl, Xacyl, $CH_2Xalkyl$;

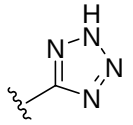
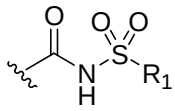
R_f , R_g and R_h are independently different or identical and are selected from Xacyl, $-CH_2Xalkyl$, $-CH_2X$-acyl and R_9,

all instances of alkyl are independently selected from Me, Et, propyl, i-propyl, butyl, iso-butyl,

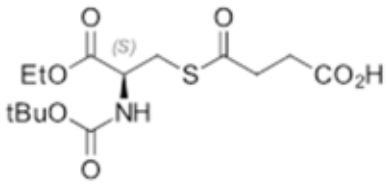
all instances of acyl are independently selected from formyl, acetyl, propionyl, isopropionyl, butyryl, tert-butyryl, pentanoyl, benzoyl,

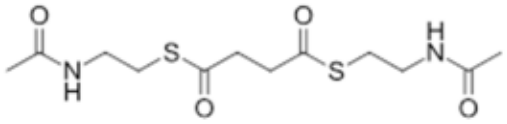
and when the dotted bond between A and B is present, the compound according to formula (I) is

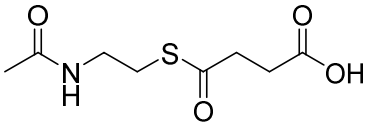
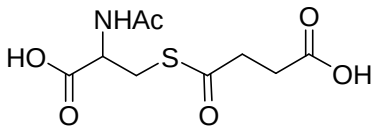


wherein X_4 is selected from –COOH, -C(=O)XR_6,  , 

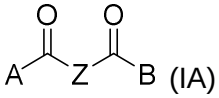
with the proviso that the compound is not



 ,

 or  .

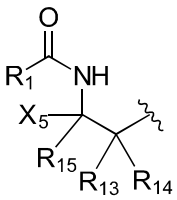
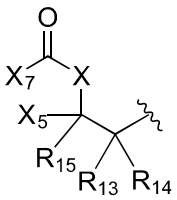
2. A compound according to claim 1 having Formula (IA)

 (IA)

or a pharmaceutically acceptable salt thereof, wherein thereof, wherein

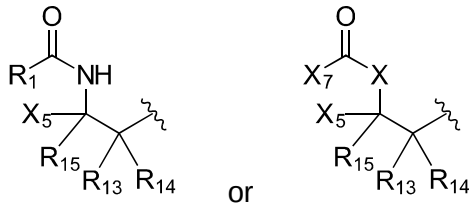
Z is –CH_2-CH_2-,

A is -SR, and R is

 or 

B is selected from -O-R', -SR''' or -OH; and

R''' is selected from one of the formulas below:



R_1 and R_3 are independently different or identical and are selected from H, Me, Et, propyl, O-Me, O-Et, O-propyl,

X is selected from O, NH, S,

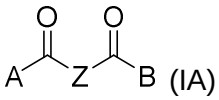
R_6 is selected from H, Me, Et,

X_5 is selected from -H, Me, Et, -COOH, $-C(=O)XR_6$, , $CONR_1R_3$

X_7 is selected from R_1, $-NR_1R_3$,

R_{13}, R_{14} and R_{15} are independently different or identical and are selected from H, Me, Et, propyl, i-propyl, butyl, iso-butyl, t-butyl, -COOH, O-acyl, O-alkyl, N-acyl, N-alkyl, Xacyl, CH_2Xalkyl, wherein alkyl and acyl are as defined herein before.

3. A compound according to claim 1 or 2 having Formula (IA)

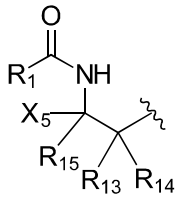


(IA)

or a pharmaceutically acceptable salt thereof, wherein

Z is $–CH_2$-CH_2-,

A is -SR and R is



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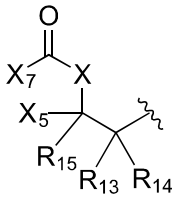
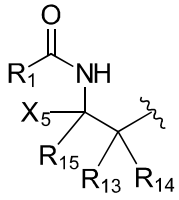


B is selected from -O-R', -SR''' or -OH; and

R''' is selected from one of the formulas below:



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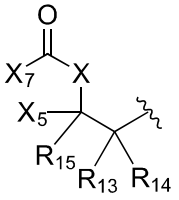


R_1 and R_3 are independently different or identical and are selected from H, Me, Et, propyl, O-Me, O-Et, O-propyl,

X is selected from O, NH, S,

R_6 is selected from H, Me, Et,

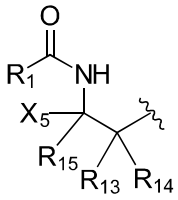
X_5 is selected from -H, Me, Et, -COOH, -C(=O)OR_6, , $CONR_1R_3$,

X_7 is selected from R_1, -NR_1R_3,

R_{13}, R_{14} and R_{15} are independently different or identical and are selected from H, Me, Et, -COOH.

4. A compound according to any one of the preceding claims, wherein B is OH or SR'''.

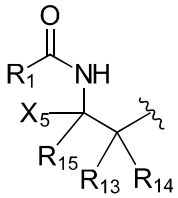
5. A compound according to any one of the preceding claims, wherein B is OH or SR''', where R''' is



.

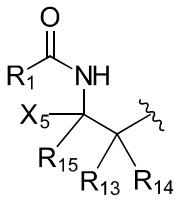
6. A compound according to any one of the preceding claims, wherein B is OH.

7. A compound according to any of the preceding claims, wherein R and/or R''' is



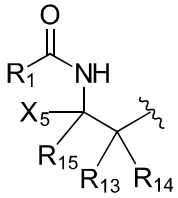
and X_5 is –H .

8. A compound according to any of claims 1-6, wherein R and/or R''' is



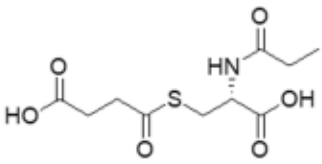
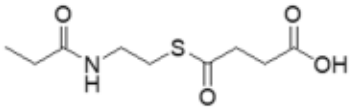
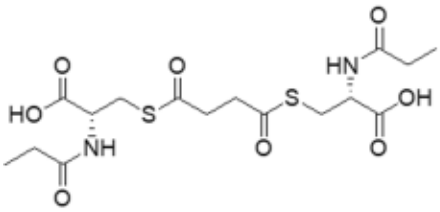
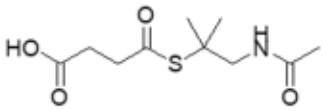
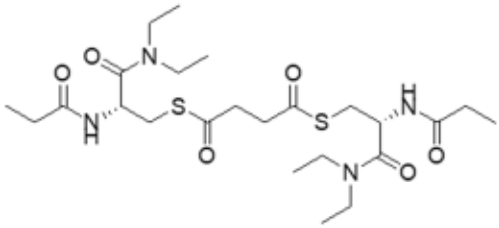
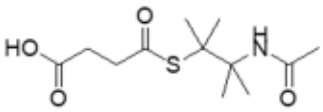
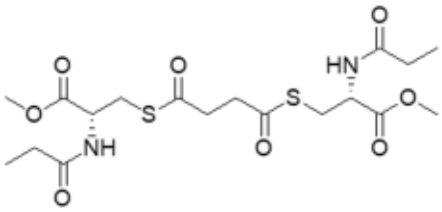
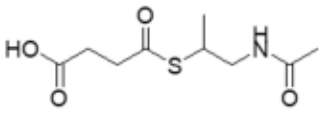
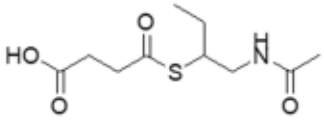
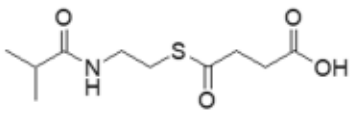
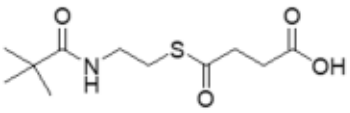
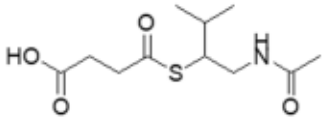
and X_5 is $COXR_6$.

9. A compound according to any of claims 1-6, wherein R and/or R''' is



and X_5 is $CONR_1R_3$.

10. A compound according to any one of the preceding claims, wherein the compound is selected from:

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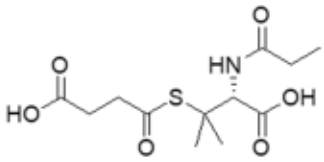

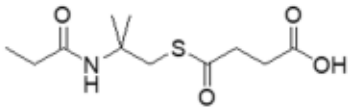

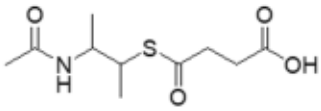

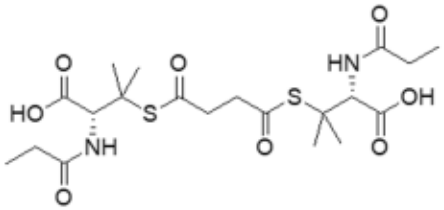

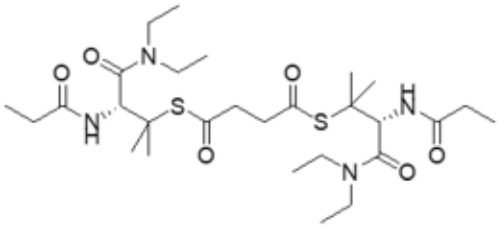

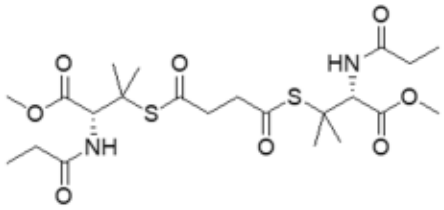

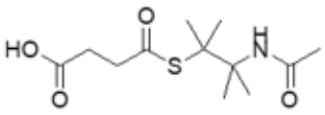

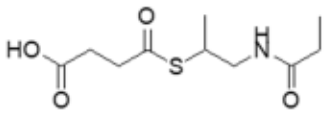

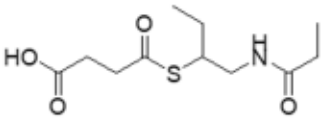

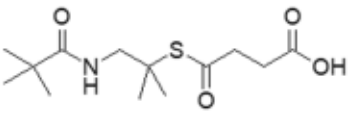

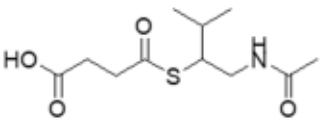

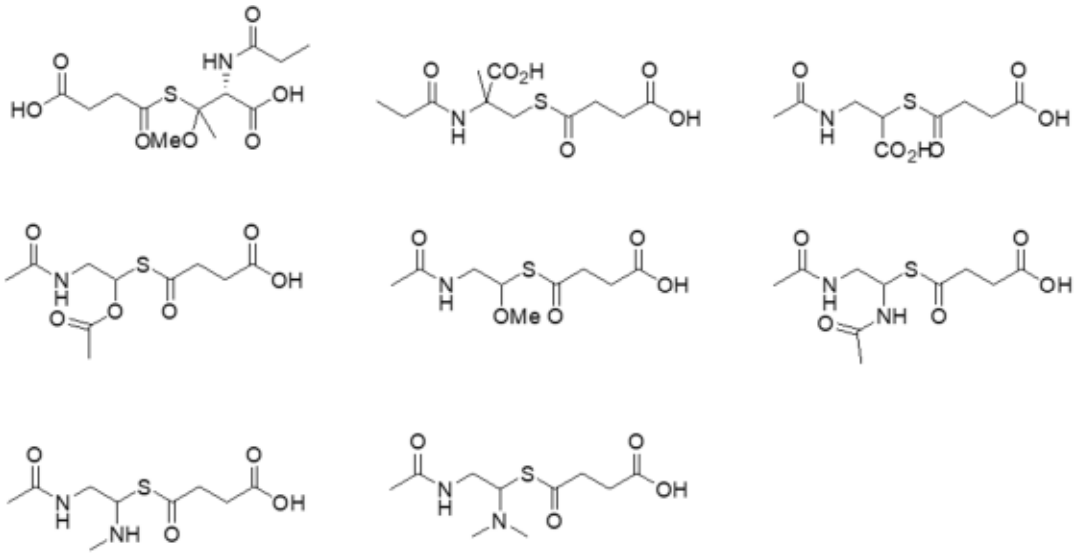

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11. A compound according to any of claims 1-10 for use in medicine.

12. A compound according to any of claims 1-10, for use in cosmetics.

13. A compound according to any of claims 1-10 for use in the treatment of diseases of mitochondrial dysfunction or disease related to mitochondrial dysfunction.

14. A compound for use according to claim 13 for prevention or treatment of mitochondrial side-effects induced by an anti-diabetic substance.

15. A composition comprising a compound of Formula (I) as defined in any of claims 1- 10 and one or more pharmaceutically or cosmetically acceptable excipients.

16. A composition according to claim 15 designed to be administered parenterally, orally, topically, via a medical device, by inhalation or via injection.

17. A compound according to any of claims 1-10 for use in the treatment or prevention of lactic acidosis.