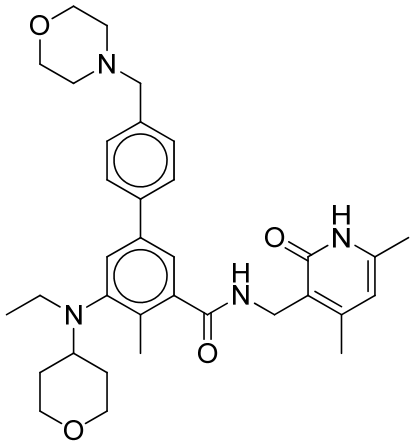
What is claimed is:

1. Use of an EZH2 inhibitor in the manufacture of a medicament for treating malignant rhabdoid tumor of the ovary (MRTO) / small cell cancer of the ovary of the hypercalcemic type (SCCOHT) in a subject, wherein the EZH2 inhibitor is



(tazemetostat), or a pharmaceutically acceptable salt thereof.

2. The use of claim 1, wherein the medicament inhibits tri-methylation of lysine 27 of histone 3 (H3K27).

3. The use of claim 1 or 2, wherein the medicament is formulated as an oral tablet.

4. The use of any one of claims 1-3, wherein the medicament is to be administered at a dose of between 10 mg/kg/day and 1600 mg/kg/day.

5. The use of any one of claims 1-3, wherein the medicament is to be administered at a dose of about 100, 200, 400, 800, or 1600 mg.

6. The use of any one of claims 1-5, wherein the medicament is to be administered twice per day (BID).

7. The use of any one of claims 1-6, wherein the SCCOHT is SMARCA4-negative.

8. The use of any one of claims 1-7, wherein the medicament is to be administered to a subject that is SMARCA4-negative.

9. The use of claim 7 or 8, wherein whether the SCCOHT or the subject is SMARCA4-negative is evaluated by a method comprising:

(a) obtaining a biological sample;

(b) contacting the biological sample or a portion thereof with an antibody that specifically binds SMARCA4; and

(c) detecting an amount of the antibody that is bound to SMARCA4.

10. The use of claim 7 or 8, wherein whether the SCCOHT or the subject is SMARCA4-negative is evaluated by a method comprising:

(a) obtaining a biological sample;

(b) sequencing at least one DNA sequence encoding a SMARCA4 protein from the biological sample or a portion thereof; and

(c) determining if the at least one DNA sequence encoding a SMARCA4 protein contains a mutation affecting the expression and/or function of the SMARCA4 protein.

11. The use of any one of claims 1-10, wherein the medicament is to be administered to a subject that is less than 40 years of age; less than 30 years of age; or less than 20 years of age.

12. The use of any one of claims 1-11, wherein the medicament is to be administered to a subject between 20 and 30 years of age, inclusive of the endpoints.

13. The use of any one of claims 1-12, wherein the medicament prevents and/or inhibits proliferation of a SCCOHT cell.

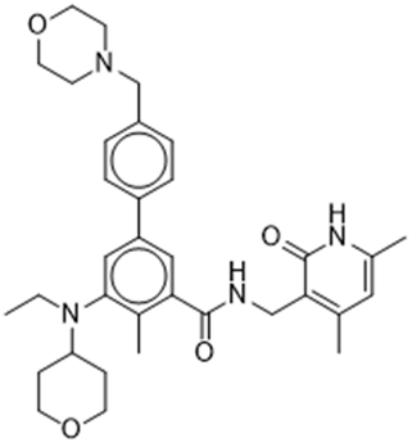
14. Use of a therapeutically effective amount of tazemetostat or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for the treatment of SCCOHT,

wherein the medicament is formulated as an oral tablet,

wherein the therapeutically effective amount is 200 mg, and

wherein the medicament is to be administered twice per day.

15. A pharmaceutical composition comprising an EZH2 inhibitor of the following formula:



(tazemetostat),

or a pharmaceutically acceptable salt thereof.

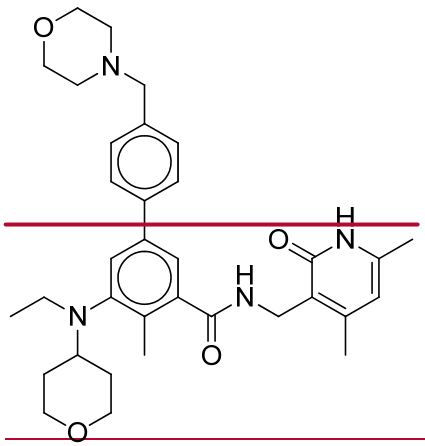
16. The pharmaceutical composition of claim 15 for use in therapy.

~~1. Use of a therapeutically-effective amount of an enhancer of a zeste homolog 2 (EZH2) inhibitor in the manufacture of a medicament for the treatment of a malignant rhabdoid tumor (MRT), optionally, wherein the MRT is INI1-negative, INI1-deficient or epithelioid sarcoma.~~

~~2. Use of a therapeutically-effective amount of an EZH2 inhibitor in the manufacture of a medicament for the treatment of malignant rhabdoid tumor of the ovary (MRTO) / small cell cancer of the ovary of the hypercalcemic type (SCCOHT).~~

~~3. The use of claim 1 or 2, wherein the EZH2 inhibitor inhibits tri-methylation of lysine 27 of histone 3 (H3K27).~~

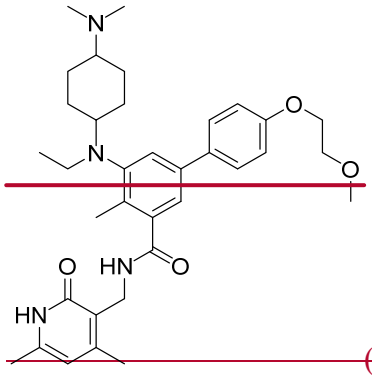
~~4. The use of any one of the foregoing claims, wherein the EZH2 inhibitor is~~



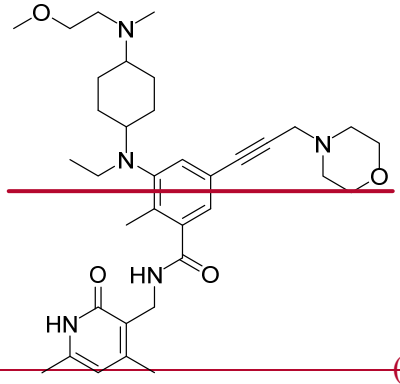
~~(tazemetostat),~~

~~or a pharmaceutically acceptable salt thereof.~~

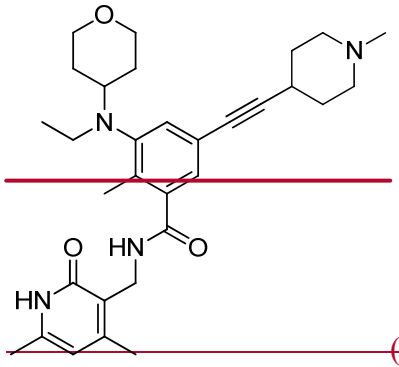
5. The use of any one of claims 1-4, wherein the EZH2 inhibitor is



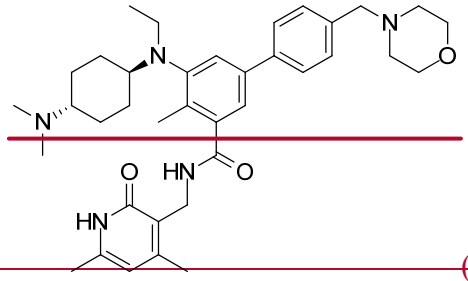
(A),



(B),



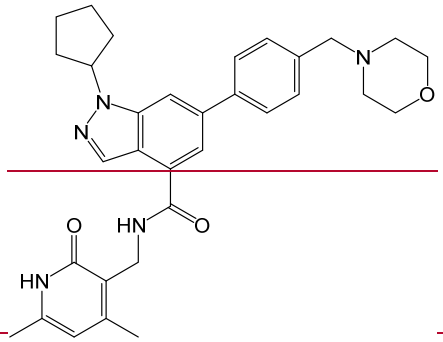
(C),



(D),

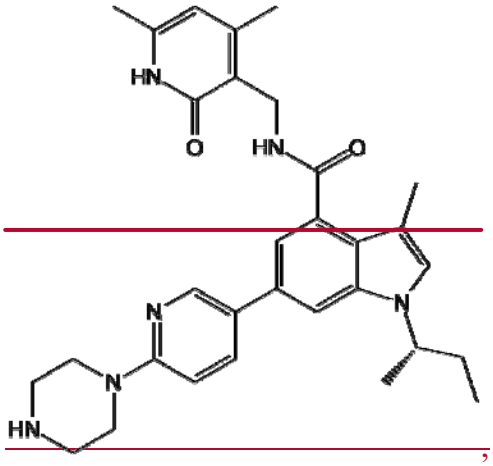
a stereoisomer, a pharmaceutically acceptable salt and/or a solvate thereof.

6. The use of any one of claims 1-4, wherein the EZH2 inhibitor is



or a pharmaceutically acceptable salt thereof.

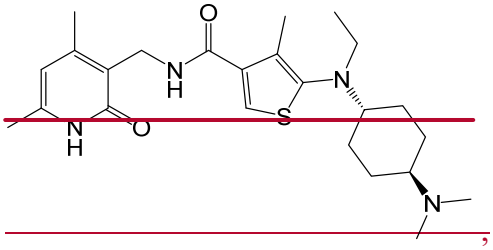
7. The use of any one of claims 1-4, wherein the EZH2 inhibitor is



,

a stereoisomer, a pharmaceutically acceptable salt and/or a solvate thereof.

8. The use of any one of claims 1-4, wherein the EZH2 inhibitor is



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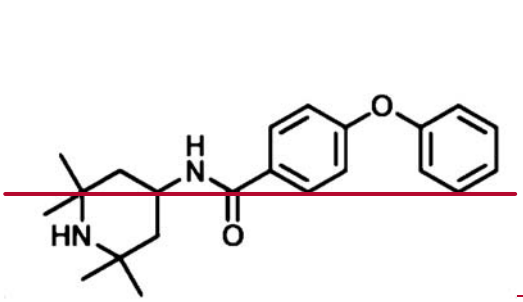
a stereoisomer, a pharmaceutically acceptable salt and/or a solvate thereof.

9. The use of any one of claims 1-4, wherein the EZH2 inhibitor is



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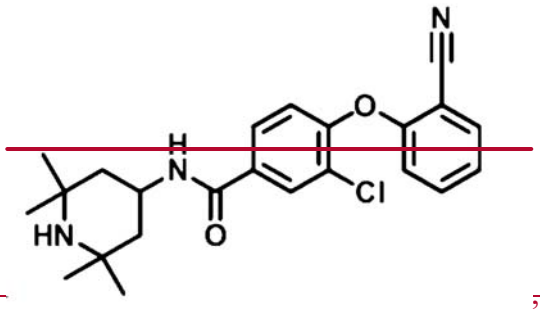

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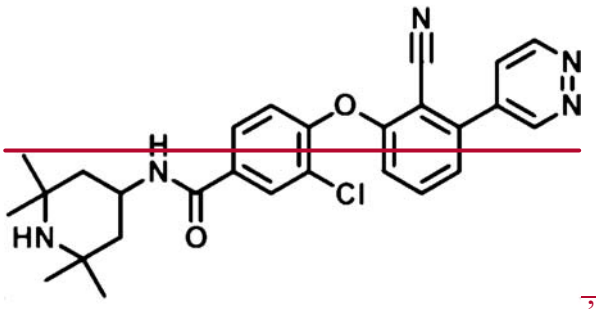

,

a stereoisomer, a pharmaceutically acceptable salt and/or a solvate thereof.

10. The use of any one of the foregoing claims, wherein the medicament is to be administered orally.

11. The use of any one of the foregoing claims, wherein the medicament is formulated as an oral tablet.

12. The use of any one of the foregoing claims, wherein the EZH2 inhibitor in the medicament is to be administered at a dose of between 10 mg/kg/day and 1600 mg/kg/day.

13. The use of claim 12, wherein the EZH2 inhibitor in the medicament is to be administered at a dose of about 100, 200, 400, 800, or 1600 mg.

14. The use of claim 12, wherein the EZH2 inhibitor in the medicament is to be administered at a dose of about 800 mg.

15. The use of any one of the foregoing claims, wherein the medicament is to be administered twice per day (BID).

16. The use of any one of claims 2-15, wherein the SCCOHT is SMARCA4-negative.

17. The use of any one of the foregoing claims, wherein the MRT is SMARCA4-negative.

18. The use of claim 16 or 17, wherein SMARCA4 expression or a function of SMARCA4 is evaluated by a method comprising:

(a) obtaining a biological sample from a subject;

(b) contacting the biological sample or a portion thereof with an antibody that specifically binds SMARCA4; and

(c) detecting an amount of the antibody that is bound to SMARCA4.

~~19. The use of any one of claims 16-18, wherein SMARCA4 expression or a function of SMARCA4 is evaluated by a method comprising:~~

~~(a) obtaining a biological sample from a subject;~~

~~(b) sequencing at least one DNA sequence encoding a SMARCA4 protein from the biological sample or a portion thereof; and~~

~~(c) determining if the at least one DNA sequence encoding a SMARCA4 protein contains a mutation affecting the expression and/or function of the SMARCA4 protein.~~

~~20. The use of claim 18 or 19, wherein the biological sample is the same biological sample used in each method of detection.~~

~~21. The use of any one of claims 18-20, wherein SMARCA4 expression or a function of SMARCA4 is evaluated by detecting an amount of the antibody that is bound to SMARCA4 and by sequencing at least one DNA sequence encoding a SMARCA4 protein.~~

~~22. The use of any one of the foregoing claims, wherein the medicament is for the treatment of a subject less than 40 years of age.~~

~~23. The use of claim 22, wherein the subject is less than 30 years of age.~~

~~24. The use of claim 23, wherein the subject is less than 20 years of age.~~

~~25. The use of any one of the foregoing claims, wherein the medicament is for the treatment of a subject between 20 and 30 years of age, inclusive of the endpoints.~~

~~26. The use of any one of the foregoing claims, wherein the treatment comprises preventing and/or inhibiting proliferation of a SCCOHT cell.~~

~~27. Use of a therapeutically-effective amount of tazemetostat in the manufacture of a medicament for the treatment of SCCOHT,~~

~~wherein the tazemetostat is formulated as an oral tablet,~~

~~wherein the therapeutically effective amount is about 800 mg/kg, and~~

~~wherein the tazemetostat is to be administered twice per day.~~