Claims

1. A compound or composition comprising said compound for use in the prophylaxis or treatment of seizures caused by one or more UBE3A mutations in a subject, wherein the one or more UBE3A mutations cause a BK channelopathy and wherein the compound or composition is an antagonist of BK channel activity.

~~2. The compound or composition of claim 1, wherein the one or more UBE3A mutations cause a BK channelopathy.~~

~~3. The compound or composition of claim 2, wherein the compound or composition is an antagonist of BK channel activity.~~

~~4.~~2. The compound or composition of ~~any one of~~ claim~~s~~ 1 ~~to 3~~, wherein the subject has Angelman syndrome or a related autism spectrum disorder.

~~5.~~3. The compound or composition of ~~any one of~~ claim~~s~~ 1 or 2~~to 4~~, wherein the compound is selected from the group comprising Paxilline, IBTX, GAL-021, small molecules, antisense molecules, aptamers, ribozymes, triplex forming molecules, small interfering RNA (siRNA), RNA interference (RNAi), and external guide sequences (EGS).

~~6.~~4. The composition of any one of claims 1 to 3~~5~~, comprising pharmaceutically acceptable salts or solvates, or pharmaceutically functional derivatives of said BK antagonist compound.

~~7.~~5. The composition of any one of claims 1 to 4~~6~~, comprising a BK channel antagonist compound with a pharmaceutically-acceptable adjuvant, diluent or carrier.

~~8.~~6. The composition of any one of claims 1 to 5~~7~~, wherein the composition is formulated for administration of a BK channel antagonist in the range of about 0.05 mg/kg to about 10 mg/kg, about 0.05 mg/kg to about 5 mg/kg, preferably about 0.3 mg/kg to about 3 mg/kg.

~~9.~~7. Use of a compound or composition for the manufacture of a medicament for the prophylaxis or treatment of seizures caused by one or more UBE3A mutations in a subject, wherein the one or more UBE3A mutations cause a BK channelopathy and wherein the compound or composition is an antagonist of BK channel activity.

~~10. The use of claim 9, wherein the one or more UBE3A mutations cause a BK channelopathy.~~

~~11. The use of claim 9 or 10, wherein the compound or composition is an antagonist of BK channel activity.~~

~~12.~~8. ~~-~~The use of ~~any one of~~ claim~~s~~ 7~~9 to 11~~, wherein the subject has Angelman syndrome or a related autism spectrum disorder.

~~13.~~9. The use of ~~any one of~~ claim~~s~~ 7 or 8~~9 to 12~~, wherein the compound is selected from the group comprising Paxilline, IBTX, GAL-021, small molecules, and functional nucleic acids such as antisense molecules, aptamers, ribozymes, triplex forming molecules, small interfering RNA (siRNA), RNA interference (RNAi), and external guide sequences (EGS).

~~14. A method of prophylaxis or treatment of seizures caused by one or more UBE3A mutations in a subject, comprising administering to a subject in need thereof a therapeutically effective amount of a compound or composition comprising an antagonist of BK channel activity.~~

~~15. The method of claim 14, wherein the one or more UBE3A mutations cause a BK channelopathy,~~

~~16. The method of claim 14 or 15, wherein the subject has Angelman syndrome or a related autism spectrum disorder.~~

~~17. The method of any one of claims 14 to 16, wherein the antagonist of BK channel activity is selected from the group comprising Paxilline, IBTX, GAL-021, small molecules, and functional nucleic acids such as antisense molecules, aptamers, ribozymes, triplex forming molecules, small interfering RNA (siRNA), RNA interference (RNAi), and external guide sequences (EGS).~~

~~18.~~10. The use~~method~~ of any one of claims 7 to 9~~14 to 17~~, wherein the composition comprises pharmaceutically acceptable salts or solvates, or pharmaceutically functional derivatives of said BK antagonist compound.

~~19.~~11. The use~~method~~ of any one of claims 7 to 10~~14 to 18~~, wherein the composition comprises a BK channel antagonist compound with a pharmaceutically-acceptable adjuvant, diluent or carrier.

~~20.~~12. The use~~method~~ of any one of claims 7 to 11~~14 to 19~~, wherein the medicament is formulated for administration of a therapeutically effective amount of the antagonist of BK channel activity is in the range of 0.05 mg/kg to

about 10 mg/kg, about 0.05 mg/kg to about 5 mg/kg, preferably about 0.3 mg/kg to about 3 mg/kg.