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 subject has Angelman syndrome or a related autism spectrum disorder.
3. The compound or composition of claim 1 or 2, 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).
4. The composition of any one of claims 1 to 3, comprising pharmaceutically acceptable salts or solvates, or pharmaceutically functional derivatives of said BK antagonist compound.
5. The composition of any one of claims 1 to 4, comprising a BK channel antagonist compound with a pharmaceutically-acceptable adjuvant, diluent or carrier.
6. The composition of any one of claims 1 to 5, 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.
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.
8. The use of claim 7, wherein the subject has Angelman syndrome or a related autism spectrum disorder.
9. The use of claim 7 or 8, 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).

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

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

12. The use of any one of claims 7 to 11, 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.