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.
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. The compound or composition of any one of claims 1 to 3, wherein the subject has Angelman syndrome or a related autism spectrum disorder.
5. The compound or composition of any one of claims 1 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. The composition of any one of claims 1 to 5, comprising pharmaceutically acceptable salts or solvates, or pharmaceutically functional derivatives of said BK antagonist compound.
7. The composition of any one of claims 1 to 6, comprising a BK channel antagonist compound with a pharmaceutically-acceptable adjuvant, diluent or carrier.
8. The composition of any one of claims 1 to 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. 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.
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. The use of any one of claims 9 to 11, wherein the subject has Angelman syndrome or a related autism spectrum disorder.
13. The use of any one of claims 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. The method of any one of claims 14 to 17, wherein the composition comprises pharmaceutically acceptable salts or solvates, or pharmaceutically functional derivatives of said BK antagonist compound.
19. The method of any one of claims 14 to 18, wherein the composition comprises a BK channel antagonist compound with a pharmaceutically-acceptable adjuvant, diluent or carrier.
20. The method of any one of claims 14 to 19, wherein the 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.