CLAIMS

WHAT IS CLAIMED IS:

1. A method of treating an ocular disease associated with one or more nonsense mutations in a subject, comprising intravitreally administering to said subject a therapeutically effective amount of one or more aminoglycosides selected from the group consisting of NB118, NB122, NB124, NB124-MeS, NB127, NB128, and NB157.

2. The method of claim 1, wherein the subject has one or more nonsense mutations selected from the group consisting of *R3X* in the *PCDH11* gene, *R245X* in the *PCDH11* gene, *R155X* in the *USH1C* gene, and *R626X* in the *USH2A* gene.

3. The method of claim 1, wherein the subject is administered about 0.3 mg/kg to about 2.5 mg/kg of the one or more aminoglycosides.

4. The method of claim 1, wherein the one or more aminoglycosides are administered by intravitreal injection.

5. The method of claim 1, wherein the one or more aminoglycosides are formulated into a pharmaceutical composition.

6. The method of claim 5, wherein the pharmaceutical composition further comprises one or more pharmaceutically acceptable carriers.

7. The method of claim 1, wherein the ocular disease associated with one or more nonsense mutations includes an inherited retinal disease, retinitis pigmentosa, Usher Syndrome, Stickler Syndrome, aniridia, Leber congenital amaurosis, and choroideremia.

8. The method of claim 1, wherein said one or more nonsense mutations are selected from the group consisting of *R3X* (*PCDH11*), *R155X* (*USH1C*), *R245X* (*PCDH15*), and *R626X* (*USH2A*).

9. A formulation for use in treating an ocular disease associated with one or more nonsense mutations in a subject, comprising one or more aminoglycosides selected from the group consisting of NB118, NB122, NB124, NB124-MeS, NB127, NB128, and NB157, wherein the formulation is for intravitreal administration.

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10. The formulation of claim 9, wherein the formulation further comprises one or more pharmaceutically acceptable carriers.

11. A kit for use in treating an ocular disease associated with one or more nonsense mutations in a subject, comprising one or more aminoglycosides selected from the group consisting of NB118, NB122, NB124, NB124-MeS, NB127, NB128, and NB157.

12. The kit of claim 11, further comprising instructions for use.

13. The method of claim 7, wherein the ocular disease associated with one or more nonsense mutations is Usher Syndrome.

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