Claims:

1. Use of acetyl-leucine, or a pharmaceutically acceptable salt thereof, in the manufacture of a medicament, for treating a neurodegenerative disease or one or more symptoms associated with a neurodegenerative disease in a subject in need thereof, wherein the neurodegenerative disease is chosen from Alzheimer's disease, amyotrophic lateral sclerosis (ALS), multiple system atrophy Parkison type (MSA-P), multiple system atrophy cerebellar type (MSA-C), frontotemporal dementia with parkinsonism, progessive supranuclear palsy, corticobasal degeneration, cerebellar downbeat nystagmus, Lewy Body dementia, and ataxia telangiectasia (Louis Barr disease), and wherein acetyl-leucine, or a pharmaceutically acceptable salt thereof is to be administered at least two times a day to achieve a total daily dose of from 0.5 g to 15 g.

2. The use according to claim 1, wherein the subject is asymptomatic.

3. The use according to claim 2, wherein the subject has been found to have a genetic and/or biochemical marker of the neurodegenerative disease.

4. The use according to claim 1, for delaying the onset of the neurodegenerative disease or one or more symptoms of a neurodegenerative disease that would otherwise be expected to manifest according to typical disease progression.

5. The use according to claim 1, wherein a therapeutically effective amount of the acetyl-leucine is to be administered to the subject in need thereof for a duration chosen from at least about 3 months, at least about 6 months, at least about 1 year, at least about 2 years, and at least about 5 years.

6. The use according to claim 1, for delaying the progression of the neurodegenerative disease or one or more symptoms associated with the neurodegenerative disease over time as compared to typical disease progression, wherein a therapeutically effective amount of the acetyl-leucine is to be administered to the subject in need thereof for a duration chosen from at least about 3 months, at least about 6 months, at least about 1 year, at least about 2 years, and at least about 5 years.

7. The use according to claim 1, for reversing the progression of the neurodegenerative disease or one or more symptoms associated with a neurodegenerative disease over time, wherein a therapeutically effective amount of the acetyl-leucine is to be administered to the

subject in need thereof for a duration chosen from at least about 3 months, at least about 6 months, at least about 1 year, at least about 2 years, and at least about 5 years.

8. The use according to claim 1, for improving in a subject in need thereof a biochemical marker of the neurodegenerative disease over time, wherein a therapeutically effective amount of the acetyl-leucine is to be administered to the subject in need thereof for a duration chosen from at least about 3 months, at least about 6 months, at least about 1 year, at least about 2 years, and at least about 5 years.

9. The use according to claim 8, wherein the biochemical marker is increased lysosomal volume.

10. The use according to any of claims 1-9, wherein a therapeutically effective amount of acetyl-leucine is to be administered to the subject in need thereof when the subject is asymptomatic.

11. The use according to claim 10, wherein the therapeutically effective amount of acetyl-leucine is to be administered after the subject has been found to have a genetic and/or biochemical marker of the neurodegenerative disease.

12. The use according to any of claims 1-4, wherein a therapeutically effective amount of the acetyl-leucine is to be administered to the subject in need thereof for a duration chosen from at least about 3 months, at least about 6 months, at least about 1 year, at least about 2 years, and at least about 5 years.

13. The use according to any of claims 1-12, wherein the acetyl-leucine is acetyl-DL-leucine.

14. The use according to any of claims 1-12 wherein the acetyl-leucine has an enantiomeric excess of the L-enantiomer or the D-enantiomer.

15. The use according to any of claims 1-4, wherein the acetyl-leucine is to be administered to the subject in need thereof at a therapeutically effect amount of from 0.5 g to 15 g per day, from 1 g to 10 g per day, from 1.5 g to 7 g per day, from 4 g to 6 g per day, or from 4 g to 5 g per day.

16. The use according to any of claims 5-12, wherein the therapeutically effective amount of acetyl-leucine is of from 0.5 g to 15 g per day, from 1 g to 10 g per day, from 1.5 g to 7 g per day, from 4 g to 6 g per day, or from 4 g to 5 g per day.

17. The use according to claim 1, for reducing the severity of the neurodegenerative disease or reducing the severity of or eliminating one or more existing symptoms associated with the neurodegenerative disease in a subject in need thereof.

18. The use according to claim 1, for providing neuroprotection in a subject having, suspected of having, or at risk of having the neurodegenerative disease, wherein a therapeutically effective amount of the acetyl-leucine is to be administered to the subject for a duration chosen from at least about 3 months, at least about 6 months, at least about 1 year, at least about 2 years, and at least about 5 years.