

Appendix: Final health state descriptions

Moderate visual impairment

Either visual acuity better than 20/200 or 6/60

Or Goldman visual field sum total degrees > 240 or radius >10 degrees

- The person has RPE65 mediated inherited retinal disease. The condition causes degeneration of the retina with associated vision loss. The person has no other significant comorbidities. They have moderate visual impairment. They can read words on a page and can cross a street safely. They may rely on a person's shape/ hair colour or voice in order to recognize them.
- The person does not need to use visual aids or a cane, but may rely on a flashlight. Young people can struggle to adapt to their progressive sight loss. It can be difficult for people to accept that they cannot drive.
- The person has difficulty with seeing at night time and during the transition (early evening). This makes it difficult to go out independently during those times. Navigating outside on their own at night can be extremely challenging, especially as they walk through areas of different light levels.
- The person can navigate through their own home with little difficulty. Going to the bathroom at night may lead them to bump into things. They may rely on additional lighting.
- The person can see the TV, but may not always recognize characters on the TV until they hear them speak.
- Navigating areas with depth – such as stairs or steps can be challenging. Near work is easier than far work, especially supported by visual aids. Cell phones/ tablets and computers can be used with little or no adaptation.
- The person has a range of career options available to them, especially if their employer offers flexibility.
- Sight loss can be extremely challenging, very difficult to come to terms with. Some people feel socially isolated partly because they find it hard to meet new people. They may be reliant on friends or services like Uber to take them to social events. They may fail to recognize someone when they walk past them. The person may live independently or may still live with parents.
- Social activities in the evening such as going to a restaurant or to the cinema can be very challenging. This may limit the enjoyment people feel from such events.
- The person can physically do sports, especially with assistance, but will struggle with sports in the evening or with sports where the ball is small. Running, swimming or gym work is possible, especially with assistance. Eye protection may be worn to prevent accidental injury.
- The person may sometimes feel low, but at other times is accepting of their vision loss. They may worry about the future.

Severe visual impairment

Either visual acuity from 20/200 to 2/500
Or Goldman visual field radius 6 to 10 degrees

- The person has RPE65 mediated inherited retinal disease. The condition causes degeneration of the retina with associated vision loss. The person has no other significant comorbidities. They have severe visual impairment. They may need a visual aid to read words on a page, or cross a street safely. They may rely on a person's shape/ hair colour or voice in order recognize them.
- The person does not want to rely on visual aids or a cane, but may rely on a flashlight. Young people can struggle to adapt to their progressive sight loss. It can be difficult for people to accept that they cannot drive.
- The person has a lot of difficulty with seeing at night-time and during the transition (early evening). This makes it difficult to go out independently during those times. Navigating outside on their own at night can be extremely challenging, especially as they walk through areas of different light levels.
- The person can navigate through their own home with little difficulty. Going to the bathroom at night may lead them to bump into things. They may rely on additional lighting.
- The person can see the TV, but may not always recognize characters on the TV until they hear them speak.
- Navigating areas with depth – such as stairs or steps can be challenging. Near work is easier than far work, especially supported by visual aids. Cell phones/ tablets and computers can be used with some adaptation.
- The person has a range of career options available to them, especially if their employer offers flexibility.
- Sight loss can be extremely challenging, very difficult to come to terms with. The person feels socially isolated partly because they find it hard to meet new people. They are reliant on friends or services like Uber to take them to social events. They may fail to recognize someone when they walk past them. The person may live independently or may still live with parents.
- Social activities in the evening such as going to a restaurant or to the cinema can be very challenging. This may limit the enjoyment people feel from such events.
- The person can physically do sports, especially with assistance, but will struggle with sports in the evening or with sports where the ball is small. Running, swimming or gym work is possible, especially with assistance. Eye protection maybe worn to prevent accidental injury.
- The person may sometimes feel low, but at other times is accepting of their vision loss. They may worry about the future.

Profound visual impairment

Either visual acuity from 20/500 to 20/1250
Or Goldman visual field radius 2 to 6 degrees

- The person has RPE65 mediated inherited retinal disease. The condition causes degeneration of the retina with associated vision loss. The person has no other significant comorbidities. They have profound visual impairment. They can't read words on a page or cross a street safely, without the help of a visual aid. They may rely on a person's shape/ hair colour or voice in order to recognize them.
- The person does not want to rely on visual aids or use a cane, which may make it harder for them to cope with the vision loss. Young people can struggle to adapt to their progressive sight loss. It can be difficult for people to accept that they cannot drive.
- The person has a lot of difficulty with seeing at night-time and during the transition (early evening). This makes it very difficult to go out independently during those times. Navigating outside on their own at night can be extremely challenging, especially as they walk through areas of different light levels.
- The person can navigate through their own home with little difficulty but may trip over things on the floor. Going to the bathroom at night may lead them to bump into things. They may rely on additional lighting.
- The person can't recognize characters on the TV until they hear them speak.
- Navigating areas with depth – such as stairs or steps can be challenging. Near work is easier than far work, especially supported by visual aids. Cell phones/ tablets and computers can be used with some adaptation such as large buttons, speak aloud buttons or voice assisted.
- The person has a range of career options available to them, especially if their employer offers flexibility, but they may end up in an entry level office job.
- Sight loss can be devastating, very difficult to come to terms with. The person feels socially isolated partly because they find it hard to meet new people. They are reliant on friends or services like Uber to take them to social events. They may fail to recognize someone when they walk past them. The person may live independently or may still live with parents.
- Social activities in the evening such as going to a restaurant or to the cinema can be very challenging. The person gets little enjoyment from such events. They have a very small group of friends.
- The person can physically do sports, especially with assistance, but will struggle with sports in the evening or with sports where the ball is small. Running, swimming or gym work is possible, especially with assistance. Eye protection may be worn to prevent accidental injury.
- The person experiences depression at times, which can be huge. Some people can experience anxiety and worry about the future.

Counting fingers

Either visual acuity 20/1250 to 20/20000

Or Goldmann visual field < 2 degrees

- The person has RPE65 mediated inherited retinal disease. The condition causes degeneration of the retina with almost complete vision loss. The person has no other significant comorbidities. Their vision is reduced to the level of counting fingers. They can't read words on a page at all. They can't cross a street safely without the help of a visual aid. They generally cannot recognize people by using shape or hair colour but instead rely on the voice.
- The person does not want to rely on visual aids but has to use a cane to navigate outside. Young people can struggle to adapt to their progressive sight loss. This person will never be able to drive.
- The person cannot see at night-time or during the transition (early evening). They can't see well during day time either, but it is much worse at night time. This makes it very difficult to go out independently, at any point in the day. Navigating outside can be extremely challenging, especially as they walk through areas of different light levels. Navigating outside is possible with support from another person or a guide dog.
- The person can navigate through their own home with little difficulty through the use of touch and hearing, but may trip over things on the floor. Going to the bathroom at night may lead them to bump into things. They may leave all of the house lights on to help them.
- The person can't recognize characters on the TV until they hear them speak.
- Navigating areas with depth – such as stairs or steps requires help. Cell phones/ tablets and computers can be used with some adaptation such as speak aloud buttons or voice assisted.
- The person is able to work but these are typically low paid roles. S/he may often just be dependent on welfare payments.
- Sight loss can be devastating, very difficult to come to terms with. People feel socially isolated partly because they can't travel independently and find it hard to meet new people. They are reliant on friends or services like Uber to take them to social events. They often fail to recognize someone when they walk past them. The person shares an apartment with other partially sighted people or lives with their parents.
- Social activities in the evening such as going to a restaurant or to meet friends can be very challenging. The person gets little enjoyment people from such events. They have a very small group of friends.
- The person cannot play any ball sports. Swimming or gym work is possible, especially with assistance. Running may be possible with a tandem partner.
- The person experiences depression at times, which can be huge. Some people can experience anxiety and worry about the future.

Hand motion to No light perception

Visual acuity worse than 20/20000

- The person has RPE65 mediated inherited retinal disease. The condition causes degeneration of the retina with complete vision loss. The person has no other significant comorbidities. They have lost all vision. They can't read words on a page at all. They can't cross a street safely without the help of a visual aid. They generally cannot recognize people by using shape or hair colour but instead rely on the voice.
- The person does not want to rely on visual aids but has to use a cane to navigate outside. Young people can struggle to adapt to their progressive sight loss. This person will never be able to drive.
- The person cannot see during the day or at night-time. This makes it very difficult to go out independently at any point in the day. Navigating outside can be extremely challenging, especially as they walk through areas of different light levels. Navigating outside is possible with support from another person or a guide dog.
- The person can navigate through their own home, but require the use of touch and hearing to do so. They may trip over things on the floor. Going to the bathroom at night may lead them to bump into things. They may leave all of the house lights on to help them.
- The person can't recognize characters on the TV until they hear them speak.
- Navigating areas with depth – such as stairs or steps requires help. Cell phones/ tablets and computers can be used if they have adaptations, adaptation such as speak aloud buttons or voice assisted.
- The person is able to work but these are typically low paid roles. S/he may often just be dependent on welfare payments.
- Sight loss can be devastating, very difficult to come to terms with. People feel socially isolated partly because they can't travel independently and find it hard to meet new people. They are reliant on friends or services like Uber to take them to social events. They often fail to recognize someone when they walk past them. The person shares an apartment with other partially sighted people or lives with their parents.
- Social activities in the evening such as going to a restaurant or to meet friends can be very challenging. The person gets little enjoyment from such events. They have a very small group of friends.
- The person cannot play any ball sports. Swimming or gym work is possible, especially with assistance. Running may be possible with a tandem partner.
- The person experiences depression at times, which can be huge. Some people can experience anxiety and worry about the future.