

Retinitis pigmentosa (RP) is a group of hereditary diseases with progressive degeneration of photoreceptors, mostly rods. RP is the main reason for blindness in young individuals, onset age of the disease varies from infancy to adulthood. RP primarily affects peripheral retina and results in a progressive constriction of the visual field. It leads to a “tunnel vision” stage, with retained central vision, and later, in the most advanced stage, to blindness. Usher syndrome involves a combination of progressive retinal degeneration in the form of RP with bilateral sensorineural hearing loss. Currently, there is no treatment for RP, however, there are some prospective therapeutic strategies ongoing, as gene therapy - process of replacing defective genes with healthy ones or replacement of photoreceptors by subretinal implants.

Sensory deprivation is a reduction or removal of stimuli delivered to the sensory organ. Vision is the most elaborated sensory input in the human brain. Approximately 20% of cortex in the human brain is dedicated to visual processing, spanning the occipital lobe and extending into temporal and parietal regions. In RP we can observe visual deprivation due to peripheral constriction of the visual field or blindness, which may affect also the visual pathway, including visual cortex. However, studies concerning this topic are scarce. In Usher syndrome there is additional sensory deprivation, hearing loss, which may additionally influence the cortex.

Over the last few decades, the development of brain imaging techniques have equipped the field of neuroscience with more sophisticated techniques to investigate the human brain including visual pathway. The vast majority of studies done so far as an extension of the diagnosis of ophthalmic diseases have been performed in magnetic field induction of 1.5 to 3T. Transferring the investigations to a higher field ensures that more accurate and specific data will be collected. Imaging with MRI in the **7T** field significantly increases the sensitivity of the magnetic resonance method and allows for extending the possibilities of already known techniques of structural, but above all functional imaging.

The purpose of the study is to investigate the effect of visual deprivation in RP on the visual pathway and brain attentional mechanism by the mean of structural and functional 7T MRI. Ophthalmological examination including visual acuity, kinetic visual field, optical coherence tomography and electroretinography will be performed in patients suffering from RP with moderate (15 patients) and advanced (15 patients) changes in the visual field due to RP, as well as 15 patients with both visual and hearing deprivation (Usher syndrome), as a control group 15 normal subjects. To exclude pathologies of the orbit and the brain all patients will undergo 1.5T routine scanning first. Next, the structural and functional examination with 7T MRI will cover the whole brain with a special emphasis on the elements of the visual pathway: from the optic nerve to the visual cortex. Later, the correlation between ophthalmological clinical features as visual field loss, retinal nerve fiber layer thickness and visual acuity and structural and functional parameters obtained with 7T MRI will be done.

High resolution 7T MRI will allow to make inferences about the mechanisms underlying differences in clinical presentation of RP. It might be also important to assess the impact of peripheral retinal loss on cortical plasticity. This may provide useful information in the context of low and high level strategies for treating different retinal diseases. Additionally, functional MRI might be a tool for assessing visual sparing, as it is more feasible and sensitive than psychophysical or ophthalmological testing.