Mitochondrial dysfunction commonly presents with ocular findings as a part of a systemic disorder. These ophthalmic manifestations can be the first sign of a mitochondrial abnormality, which highlights the key role of a comprehensive ophthalmic assessment. On the other hand, a number of visually disabling genetic and acquired eye diseases with no curative treatment show abnormal mitochondrial function.

Recent advances in mitochondrial research have improved our understanding of previously unexplained ocular disorders utilising better diagnostic approaches. Further studies on mitochondrial dysfunction and novel modalities of treatment will help to improve outcomes of these conditions. In this review article we discuss the clinical picture of common mitochondrial-related eye diseases, diagnostic approaches and possible treatment options including a very recent interesting report about gene therapy, with a particular focus on glaucoma.

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