1. Background
Mitochondria have challenged the physicians and cell biologists for the last 50 years. They are one of the most complex organelles within the human body and are responsible for regulating a variety of functions from metabolism to cell death and are associated with a number of diseases.
A study of mitochondrial diseases/encephalomyopathies is relevant to Indian population which is subdivided into a number of castes, sub-castes and tribes, each of which form endogamous communities. Almost all mitochondrial diseases have neuromuscular involvement. Deficiency of OXPHOS is the commonest cause of neuromuscular disorders in childhood. Among the OXPHOS deficiencies, complex I deficiency is frequently accounting for about 30% of mitochondrial disorders. Mutations in nuclear or mtDNA encoded structural subunits of respiratory chain are the commonest cause of these disorders at genetic level. Identification of these mutations can help us to understand the molecular background of OXPHOS deficiency. Further consequences of OXPHOS deficiency upon cell function in terms of ROS levels and apoptosis will help in better diagnosis and to develop treatment strategies. Any efforts to define the origin at genetic and biochemical level will be of value to families with affected children and to design therapies.

2. Objectives
The specific objectives of this research were:
1. Biochemical analysis of the mitochondrial respiratory chain defects in children with possible mitochondrial encephalomyopathies.
2. To know the genetic background of respiratory chain or OXPHOS defect at mitochondrial genome level.
3. To check the status of reactive oxygen species (ROS) and antioxidant defenses in cell lines established from encephalomyopathic patients.