***Abstract**

Background: Muscle biopsy is used for evaluating morphologic, biochemical and ultrastructure analysis of the muscles which help to reach specific neuromuscular diagnosis. However, muscle biopsy is an invasive procedure associated with high cost, time-consumption, resource-demanding and involves slight risk to the children. We aimed to identify the yield of muscle biopsy in children suspected to have neuromuscular diseases.

Methods: We retrospectively reviewed muscle biopsy in these children in our institution between 2004 and 2014. Clinical presentations, complication of biopsy, pathological results and change in management decision were reviewed and analyzed.

Results: Of 92 patients, 61 (66.3%) were male. Mean age at biopsy was 7.1 years. Clinical presentations included weakness (90.2%), gross motor developmental delay (31.5%), abnormal gait (26.1%), hypotonia (21.7%), ptosis (5.4%), respiratory failure (3.3%), and seizure (2.2%). There were no perioperative complications. A definite diagnostic finding can be made in 74 patients (80.3%). Biopsy resulted in changing in management decision in 87% including genetic and prognosis counselling, medication treatment and surveillance for complication.

Conclusions: Muscle biopsy is useful and safe in children suspected to have neuromuscular diseases especially muscle diseases. The definite pathologic diagnosis also helps physician for further management including medication treatment, genetic counselling, surveillance for complication, and prognosis.