Myotonic dystrophy type 2 (DM2) is one of the most common forms of muscle dystrophies in adults and remains incurable. It is autosomal dominant disease associated with expansional instability of CCTG tetranucleotide repeats in the first intron of the cellular retroviral nucleic acid binding protein (CNBP) gene. While normal individuals have up to 26 CCTG repeats, in DM2 patients the repeats expand beyond several thousands of copies. Pathogenesis of DM2 is linked to expression and abnormal processing of mutant CNBP pre-mRNA and involves RNA toxic gain-of-function effect. The mutation-containing intron 1 (*i1CCUG*) escapes rapid degradation and instead accumulates in the nucleus as stable foci. Their presence is widely considered a major source of abnormalities in DM2 cells and a trigger of pathogenesis. The effect of CCTG expansion on CNBP expression is currently controversial, with some studies reporting no effect, and other studies a decrease in CNBP mRNA and protein levels in DM2 cells and tissues. The most recent results based on high-throughput RNA-seq analysis have found aberrant splicing of the host intron 1 and its retention was detected in DM2 when compared with non-DM2 controls. The CCTG expansion is located in a large 12k nt intron which is placed in the 5'UTR of *CNBP*. In mutant allele the i1 length vary significantly due to unstable nature of CCTG repeat, and in some cases it may reach over 50k nt. Conventional model of splicing suggests that nearly all introns are removed as a single unit via one cycle of spliceosome assembly and splicing. However, examples of intron excision via multiple recursive splicing (RS) events have been described and as recently reported in human genes most introns are removed from pre-mRNAs in smaller multiple pieces rather than spliced as whole units in one step reaction. This process results in the generation of transient splicing intermediates that are source of a final mRNA. Aberrant processing of these precursors affecting their fate may be a source of pathogenesis. In this project we aim to test the hypothesis that large i1 of CNBP is spliced via non-canonical multiple RS events via un-annotated splice sites. The recognition of RS sites by the spliceosome apparatus is perturbed in DM2 due to functional depletion of splicing factors reported in patients and evidenced as global aberrant alternative splicing. This in turn causes i1 retention and its aberrant turnover detected in DM2 cells and tissues as nuclear RNA foci. To test our hypothesis we will: **(i)** analyze CNBP RNA processing pattern in vivo using metabolic labeling of nascent RNA with 4sU in different cell types from DM2 and non-DM2 controls; the nanopore sequencing will be performed to determine the extent of RS in these cells; **(ii)** determine the category of i1 splicing by-products by RNA-seq analysis of lariat and circular RNAs; **(iii)** using different tissues from DM2 patients and unaffected controls we will validate the results obtained in cultured cells; **(iv)** determine the global extent of RS in human cells and tissues of DM2 in comparison to controls and their prospective function in DM2 pathogenesis.