

Supplementary Table 1: Pathology domains and scoring/evaluation criteria used for muscle biopsy assessment

Pathology domain	Score	Evaluation criteria
Muscle fibre domain	0~6	
Perifascicular atrophy	0,1,2	Absent - score 0. Present in one or 2 fascicles - score 1. Present in 3 or more fascicles - score 2.
Necrosis	0,1,2	Absent - score 0. < 5% fibres- score 0. > 5% fibres- score 2.
MHC Class I over- expression	0,1,2	< 1% area of muscle tissue - score 0. Between 1% and 20% area of muscle tissue - score 2. ≥20% area of muscle tissue - score 2.
Inflammatory domain	0~4	
CD3+ cell infiltration*	0,1,2	None, or less than 4 cells in × 25 field - score 0. > 4 cells in × 25 field or 1 cluster - score 1. >2 clusters or diffusely infiltrating cells - score 2.
CD20+ cell infiltration*	0,1,2	
Vascular domain	0,1	Mural thickening and/or endothelial swelling and/or transmural inflammation in arteries/arterioles. Absence –score 0; Presence – score 1.
Connective tissue domain	0,1	Fibrosis in each of endomysial and perimysial distributions. Absence –score 0; Presence – score 1.

\*Area of endomysial, perimysial and perivascular; Cluster infiltration: > 10 cells in ×25 field; Diffuse infiltration > 25 cells in×25 field.