

Supplementary Information

Very long-/ and long Chain-3-Hydroxy Acyl CoA Dehydrogenase Deficiency correlates with deregulation of the mitochondrial fusion/fission machinery

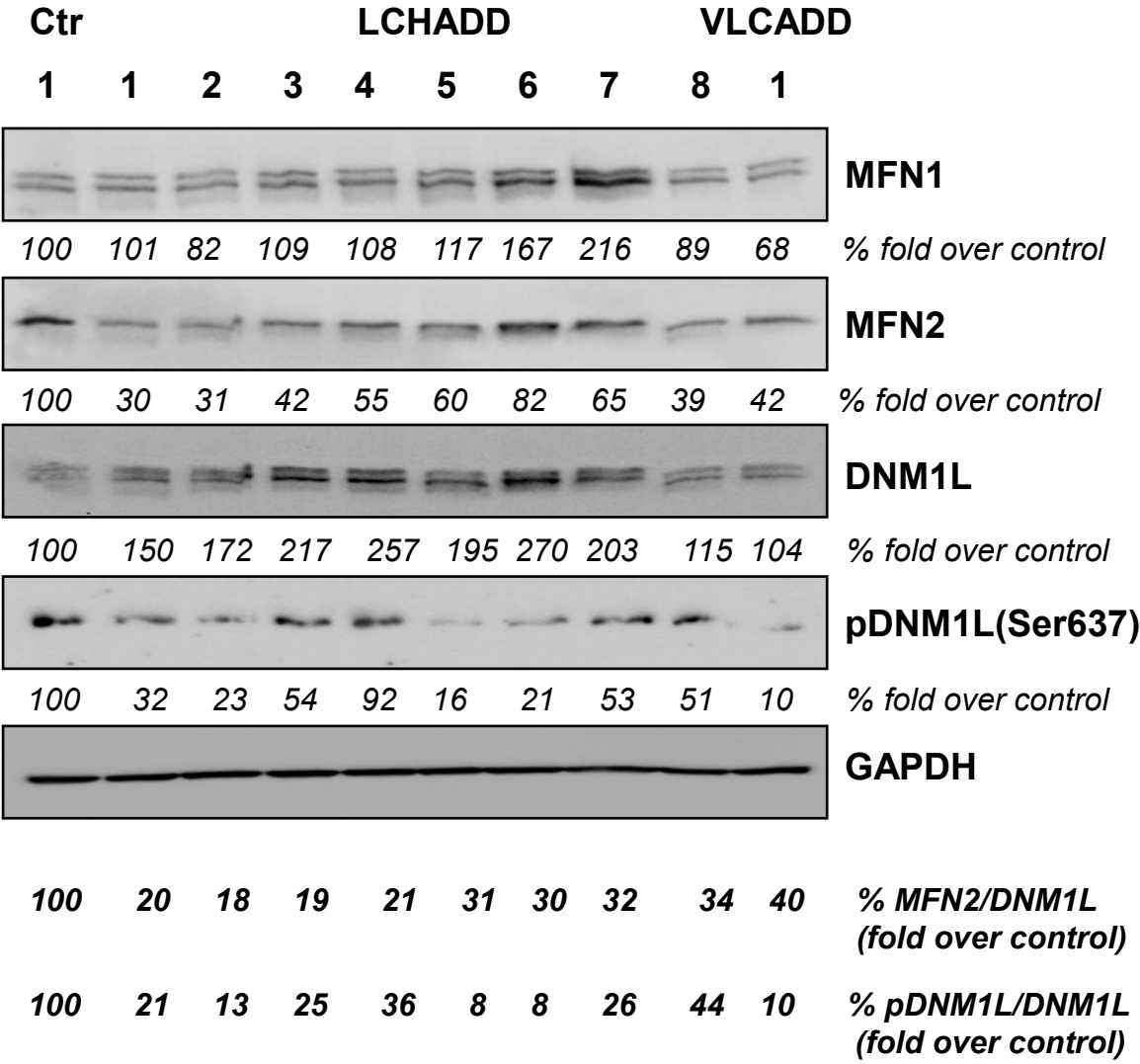
Judith Hagenbuchner¹, Sabine Scholl-Bürigi², Daniela Karall^{2*}, and Michael J. Ausserlechner^{2*}

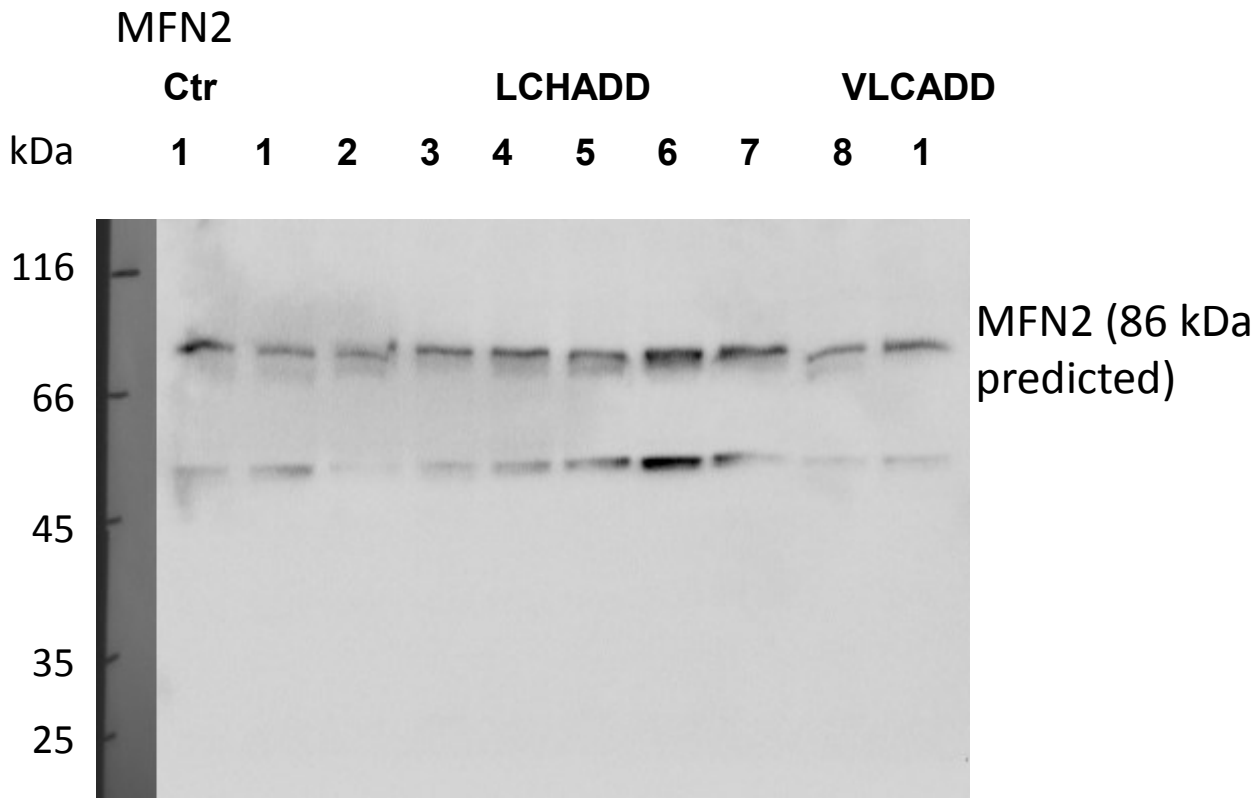
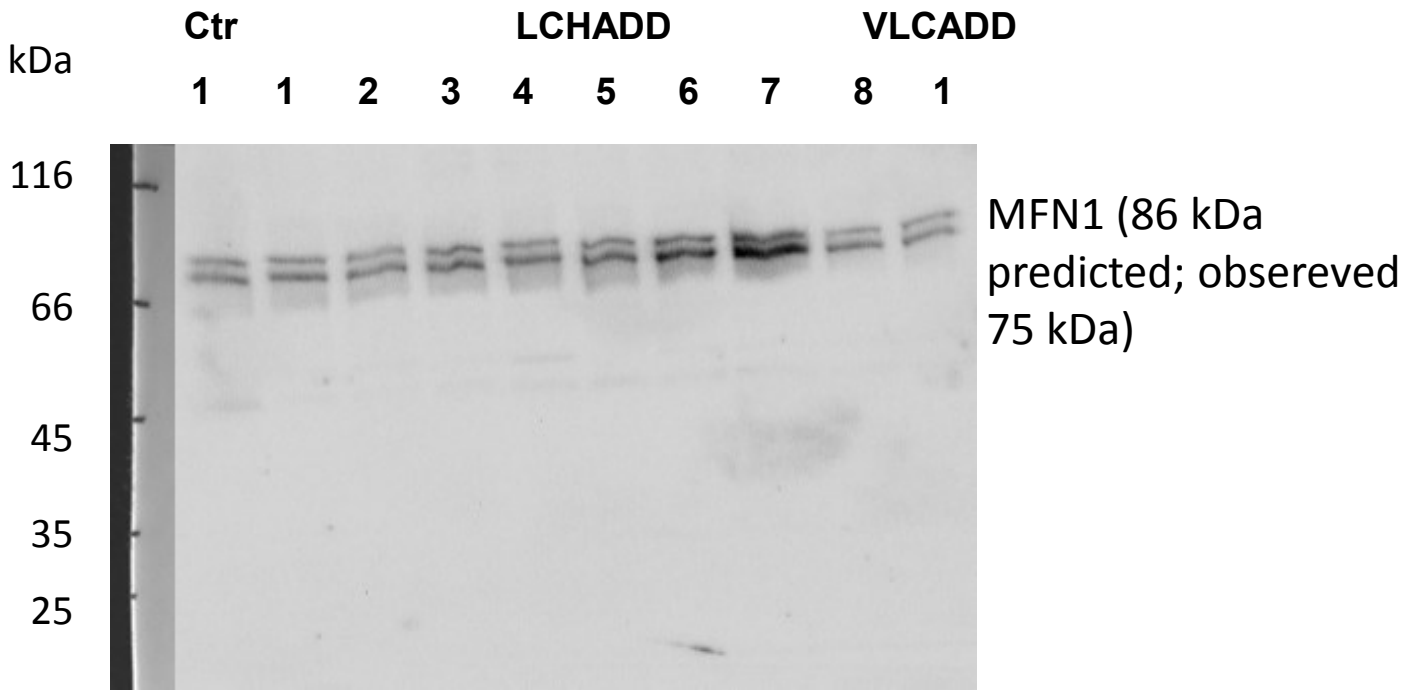
¹Departments of Pediatrics II and ²Pediatrics I, Medical University Innsbruck, Innsbruck, Austria

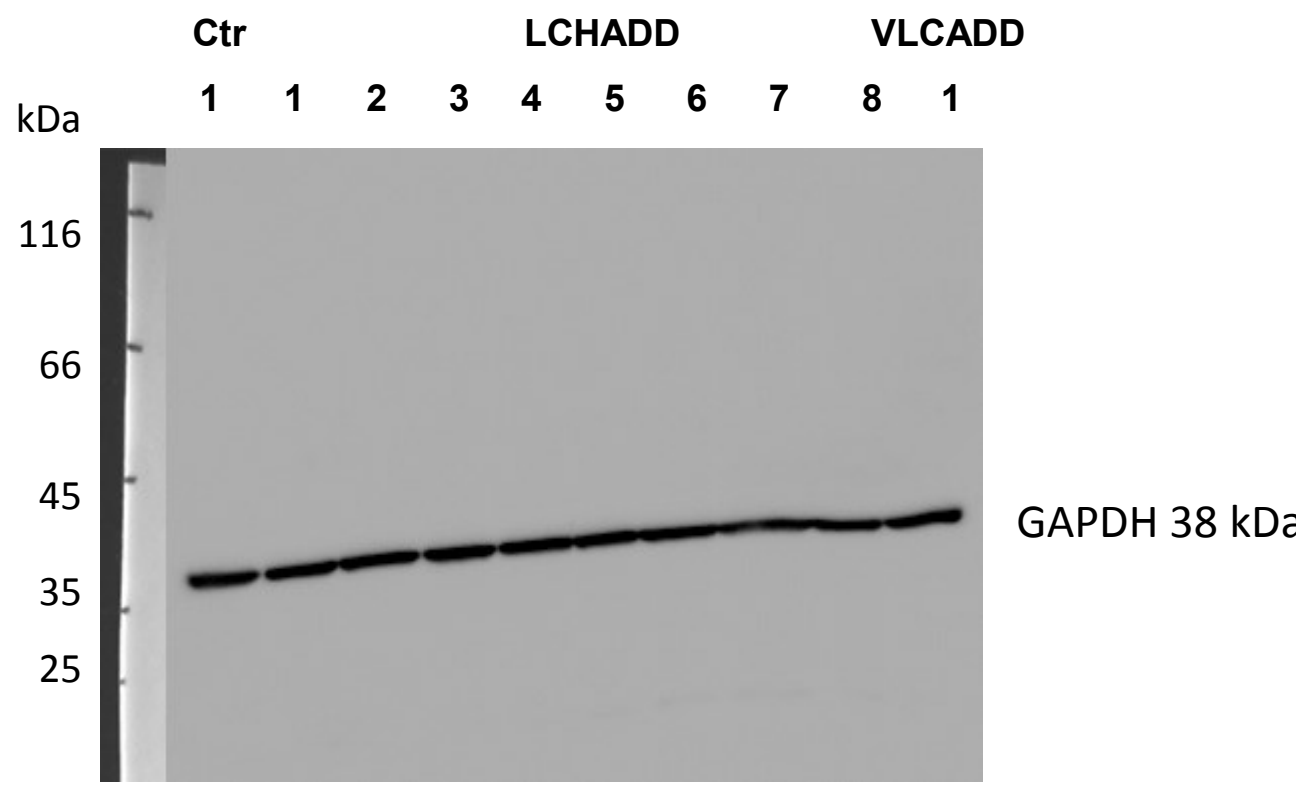
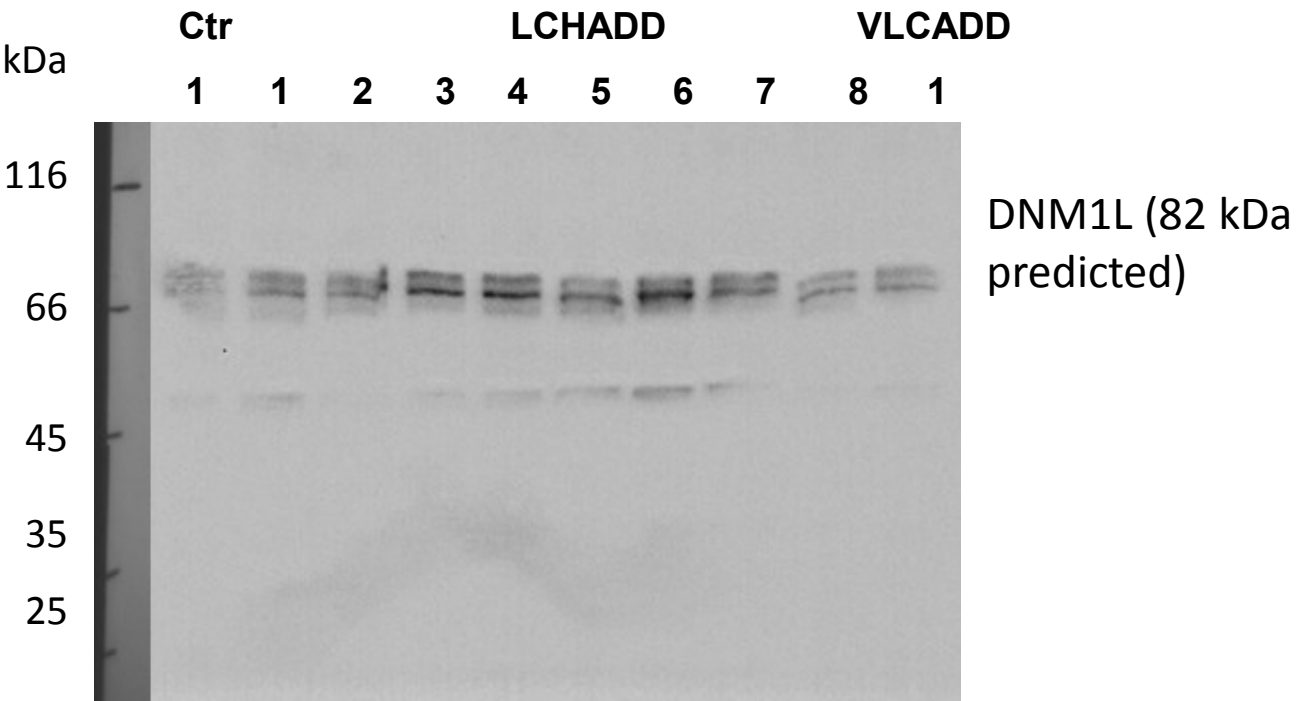
*corresponding authors

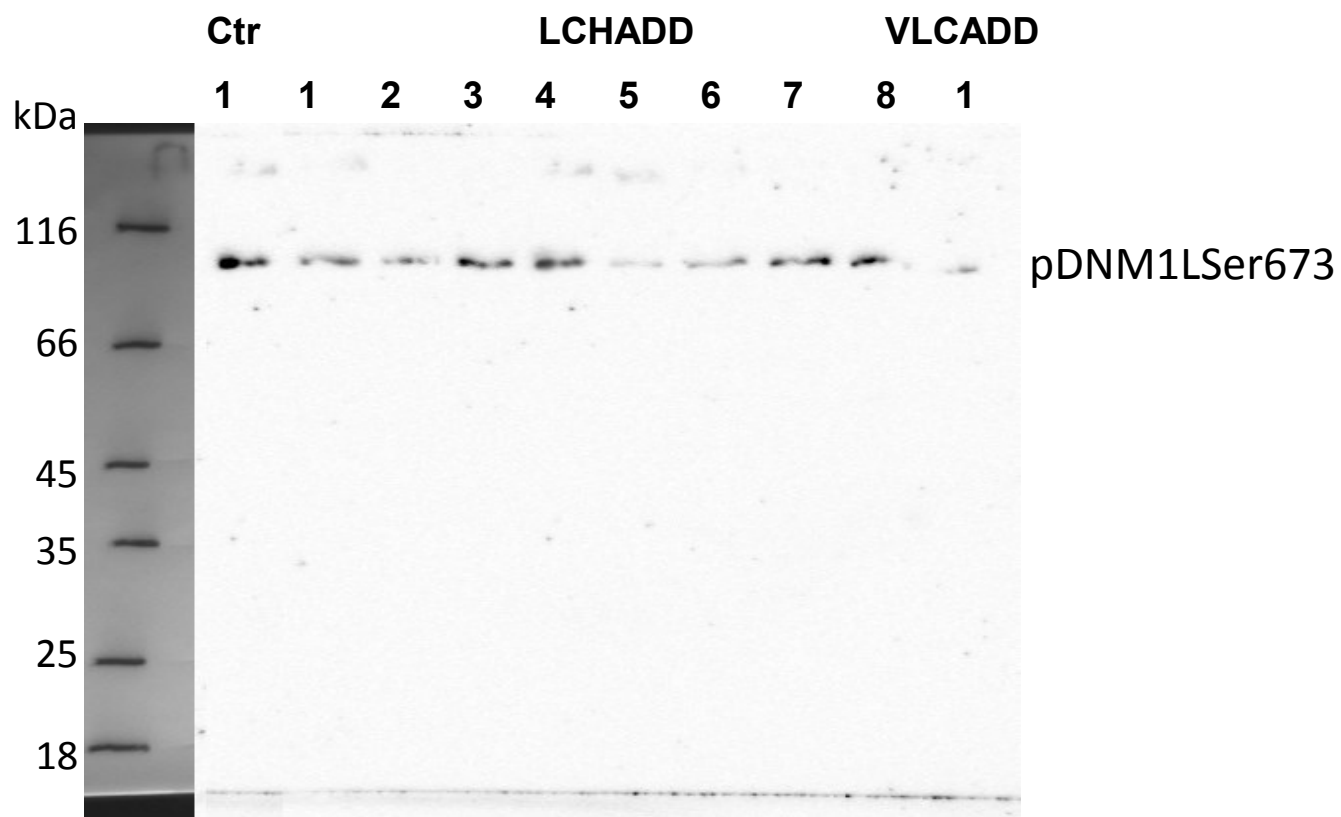
Supplemental Figure 1: Original uncropped blots

**A) Analyses of Fusion/Fission proteins in LCHADD/VLCADD patients
PANEL 1:**

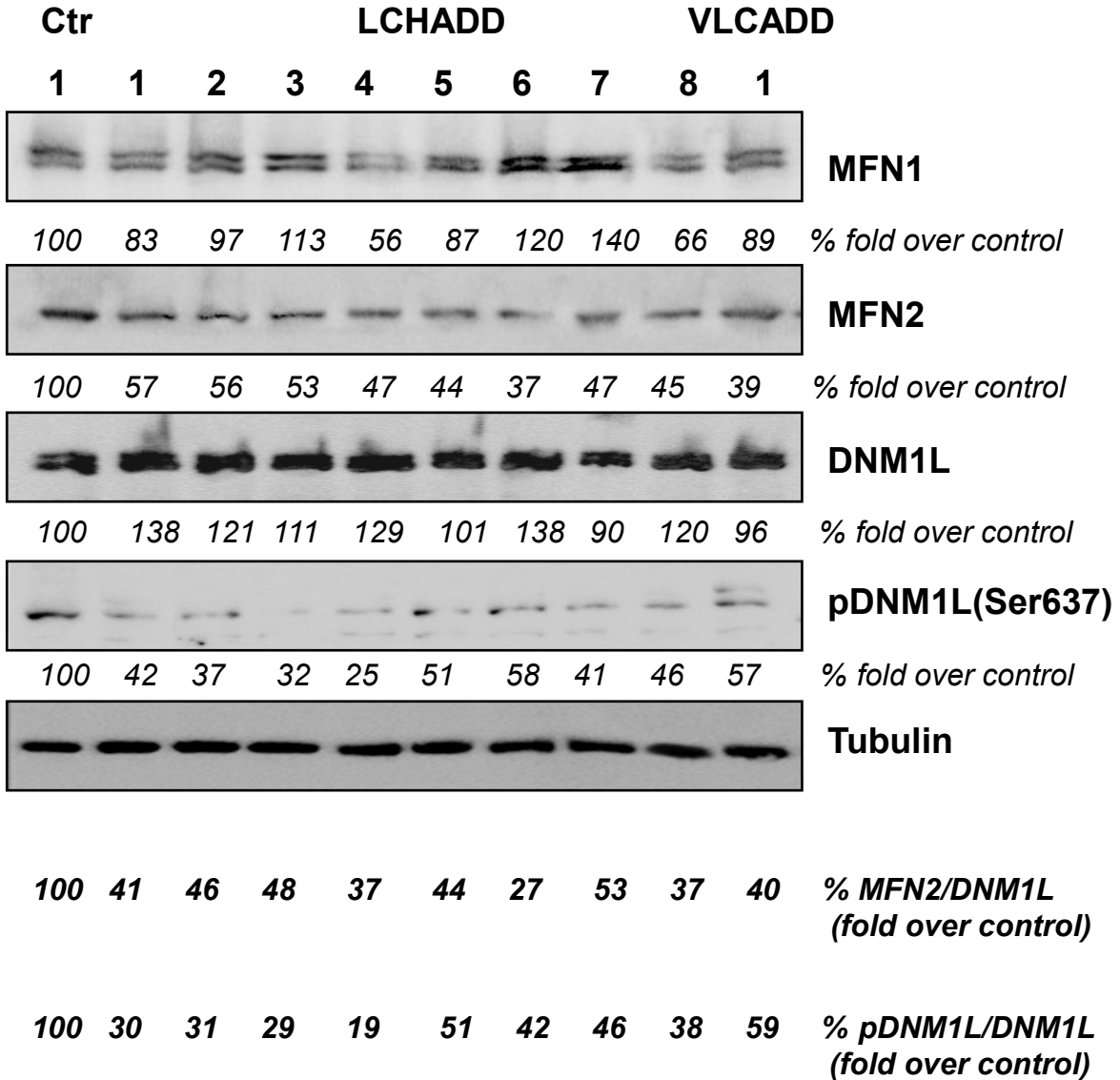


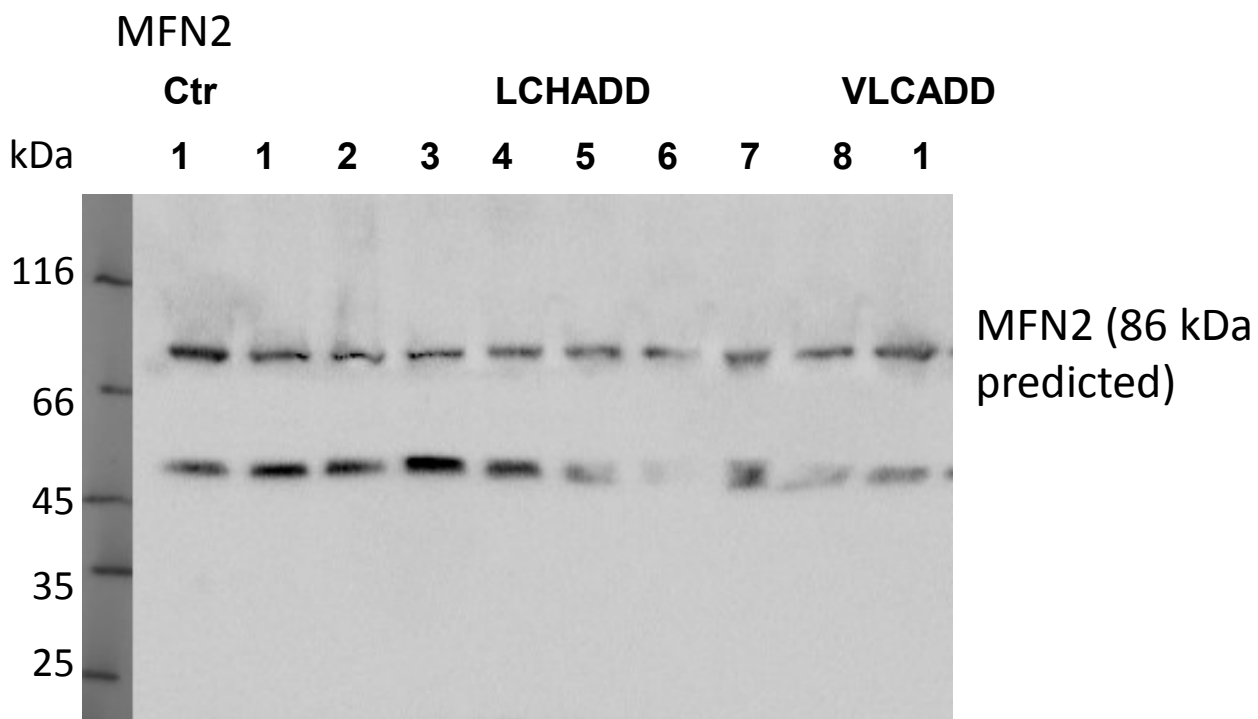
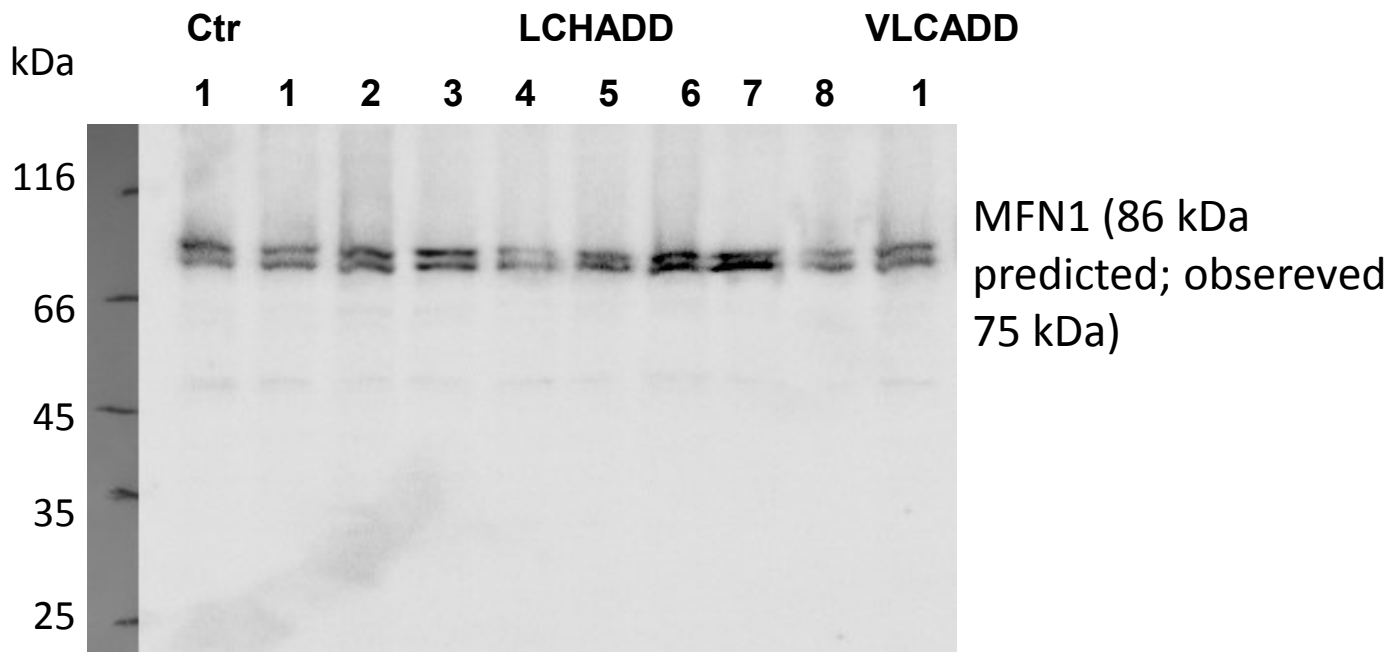


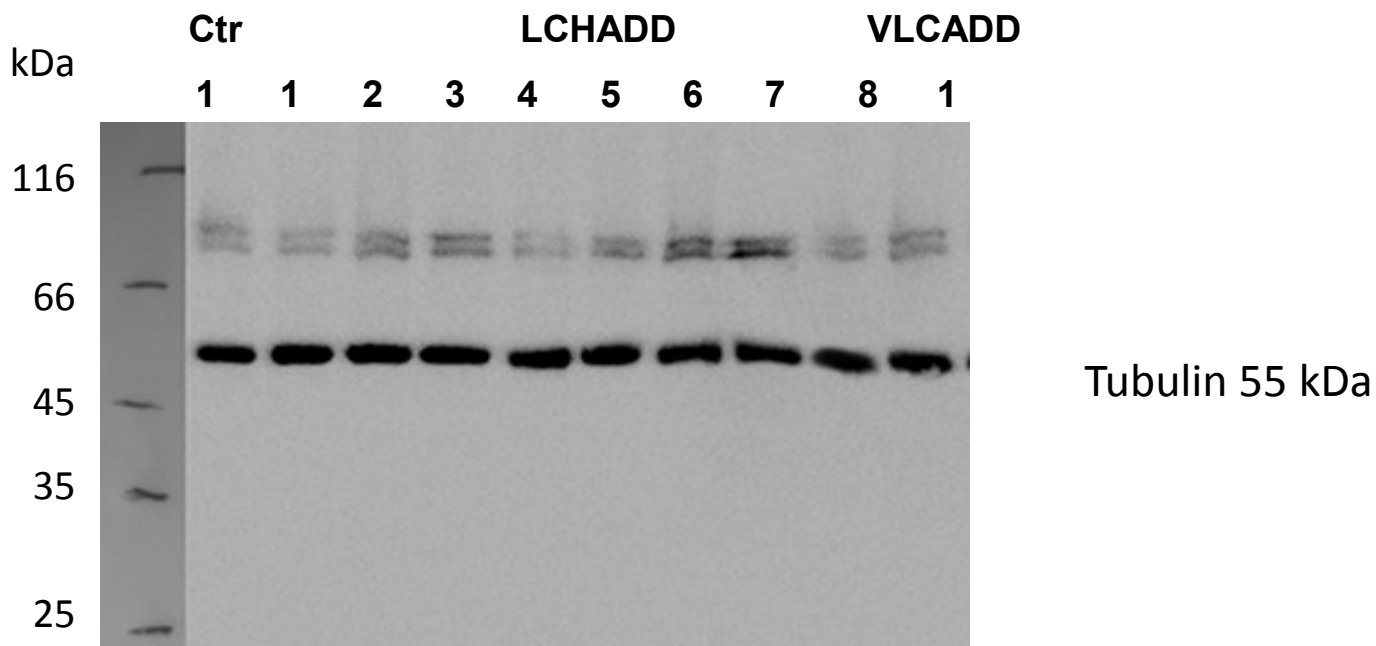
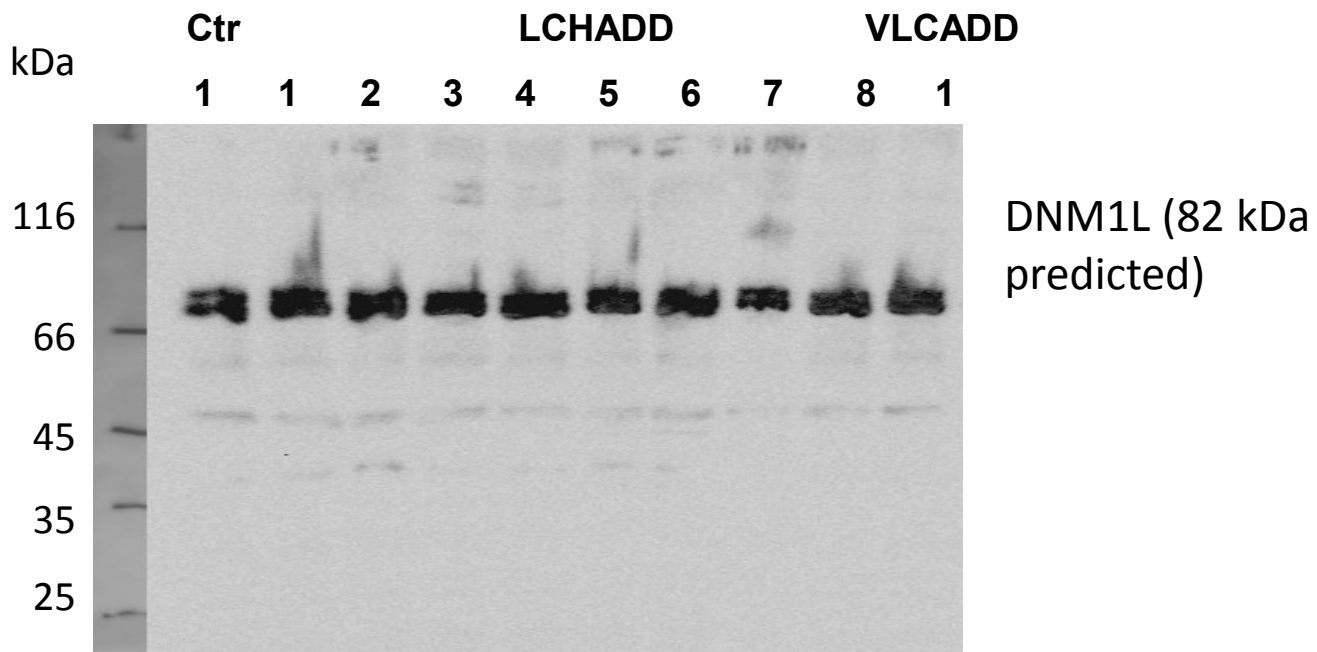




**Analyses of Fusion/Fission proteins in LCHADD/VLCADD patients
PANEL 2:**







Ctrl

LCHADD

VLCADD

1

1

2

3

4

5

6

7

8

1

kDa

116

66

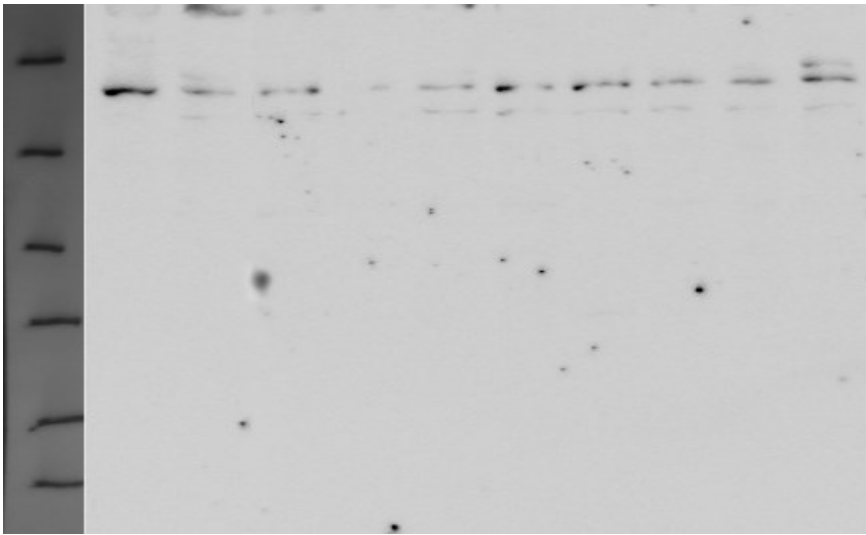
45

35

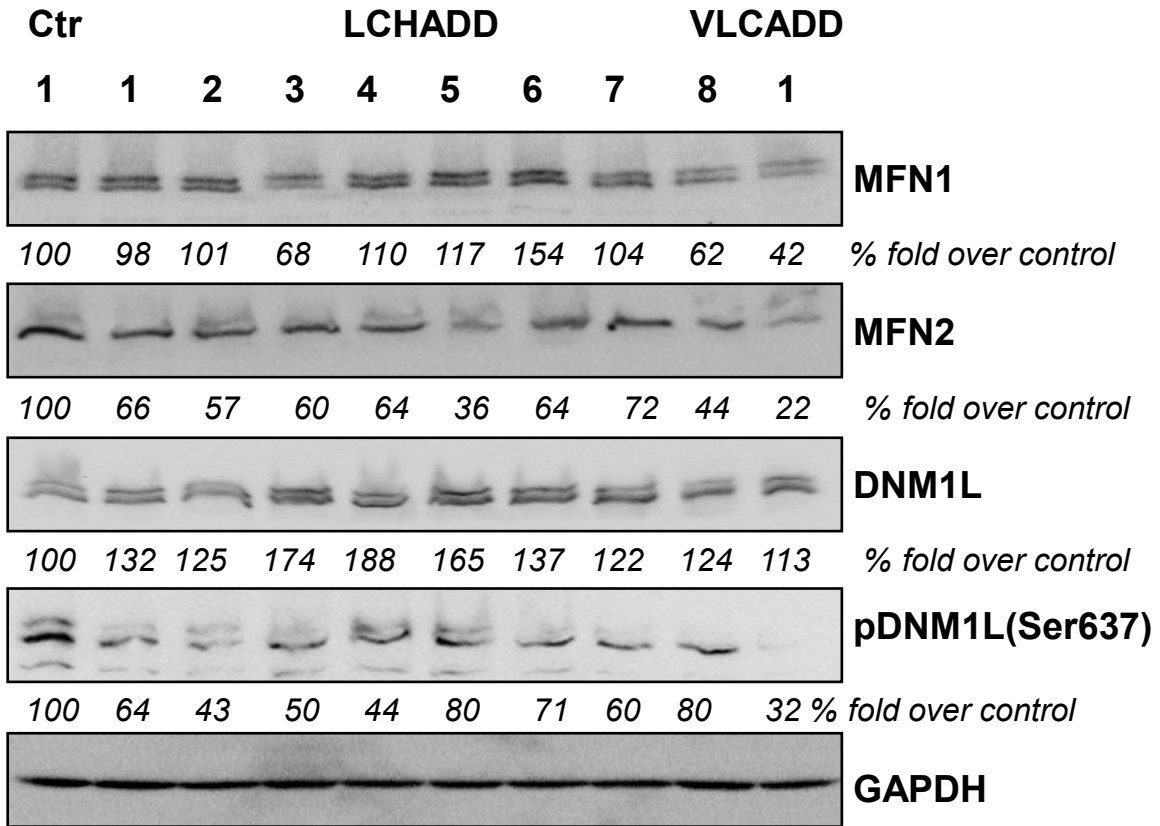
25

18

pDNM1LSer673

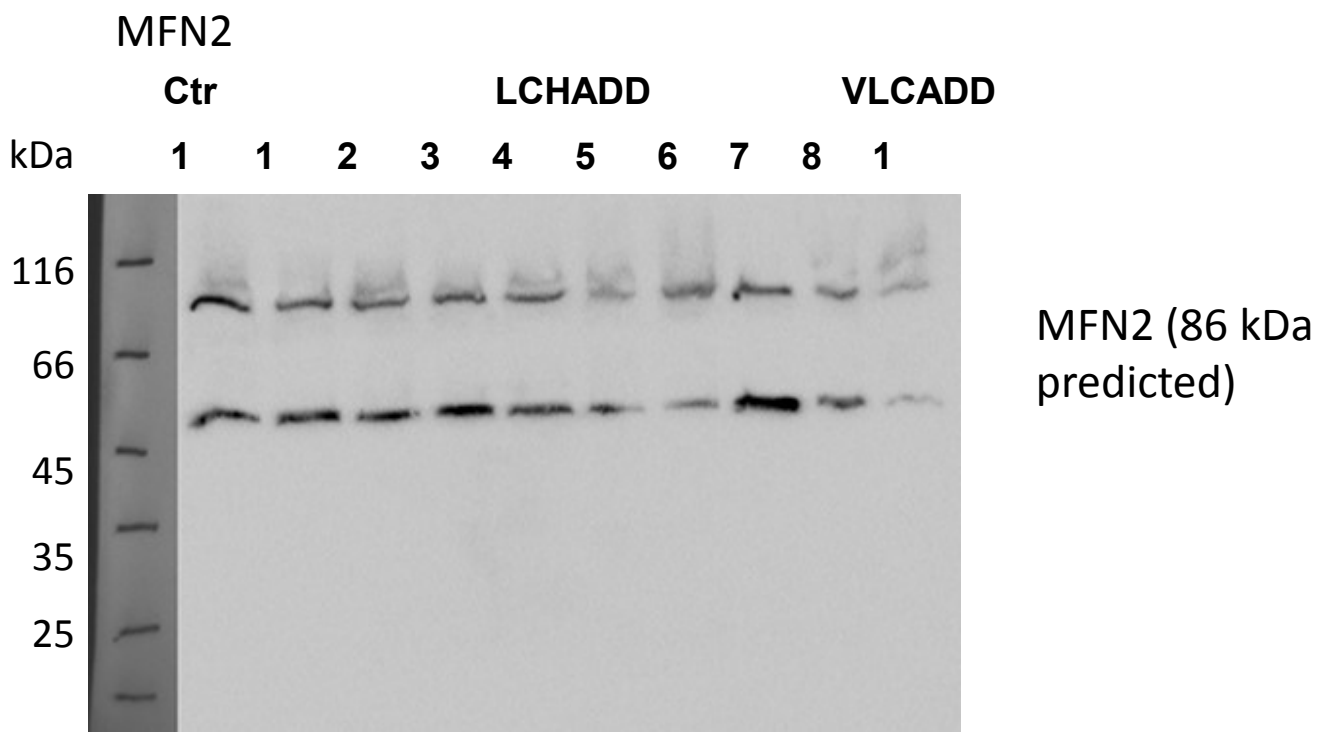
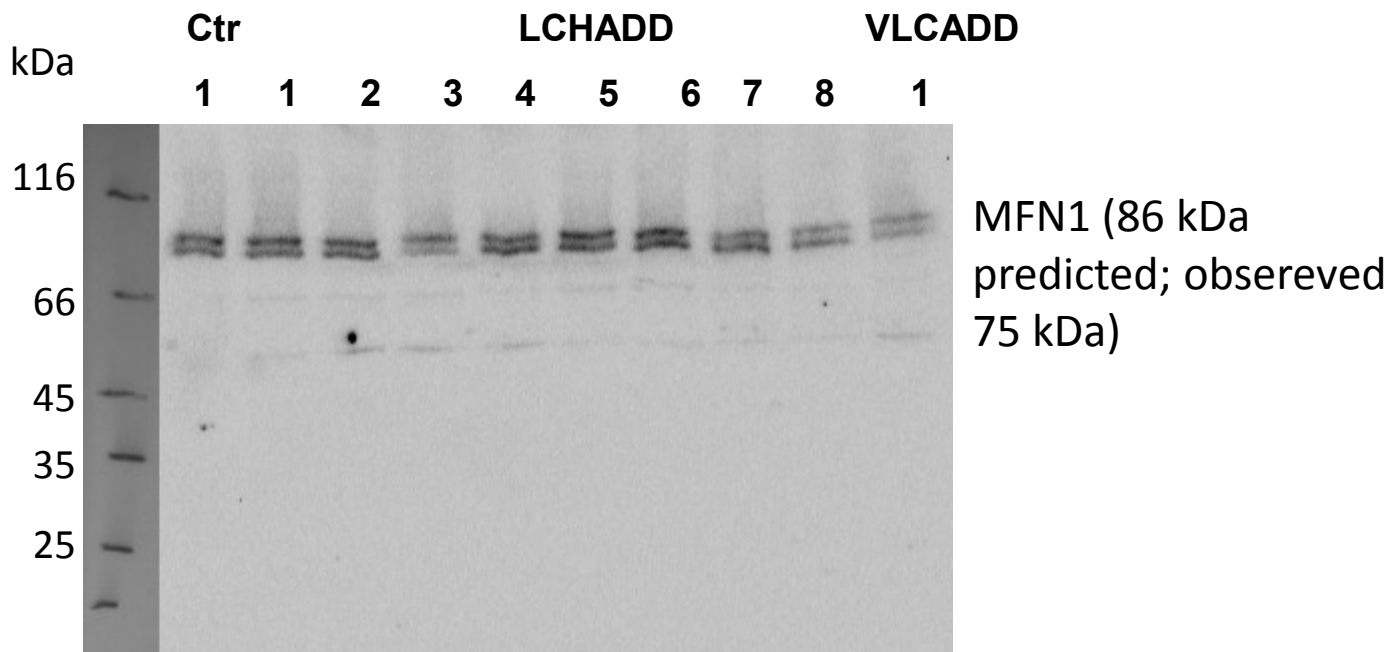


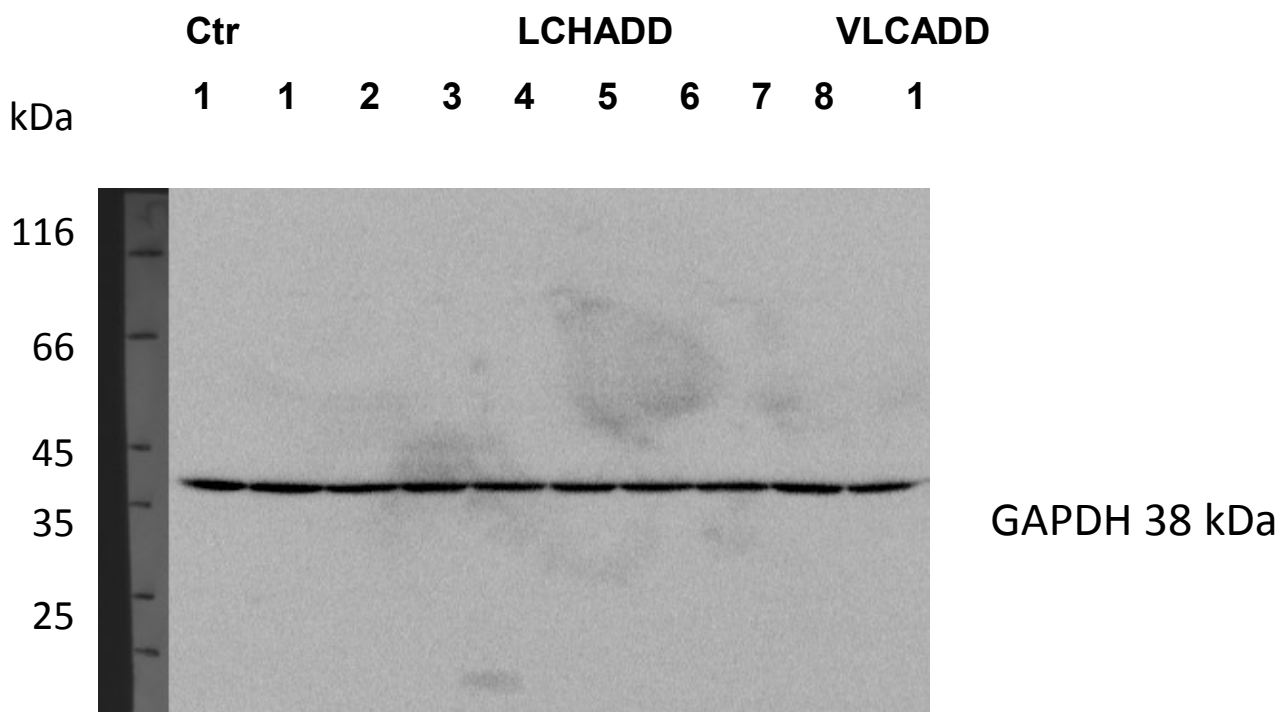
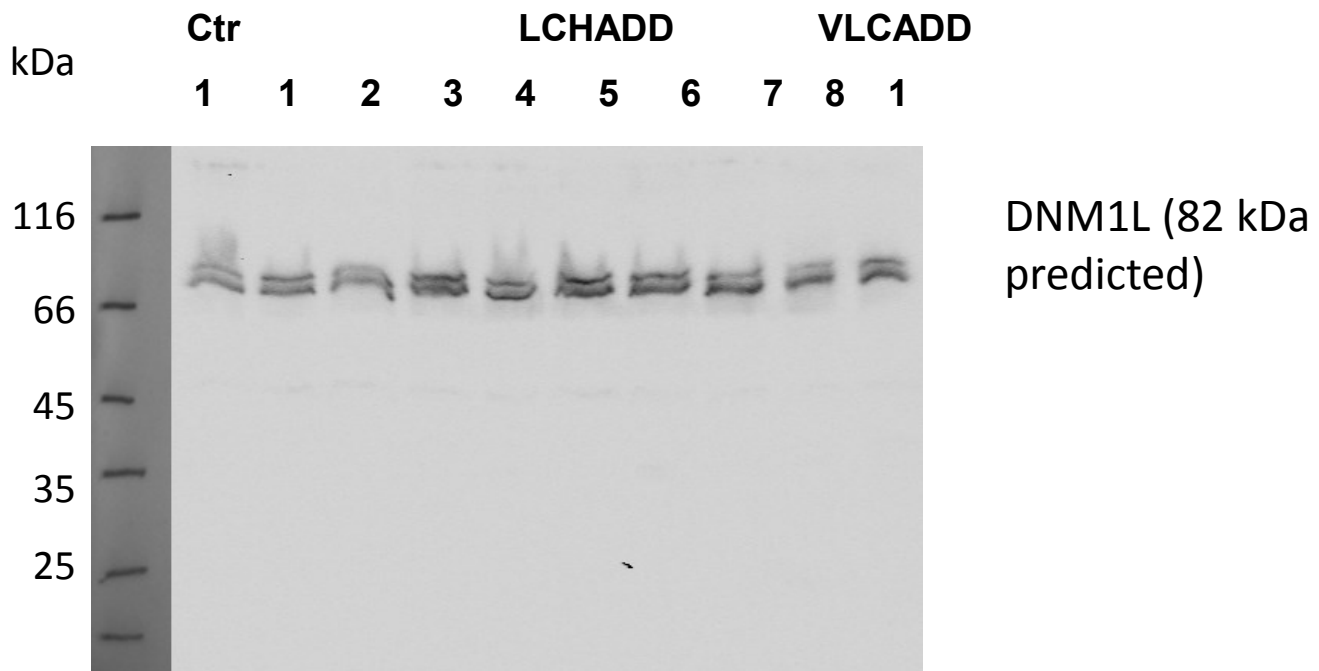
**Analyses of Fusion/Fission proteins in LCHADD/VLCADD patients
PANEL 3:**



100 50 45 35 29 22 47 60 36 20 % MFN2/DNM1L
(fold over control)

100 48 34 29 24 48 52 49 64 28 % pDNM1L/DNM1L
(fold over control)





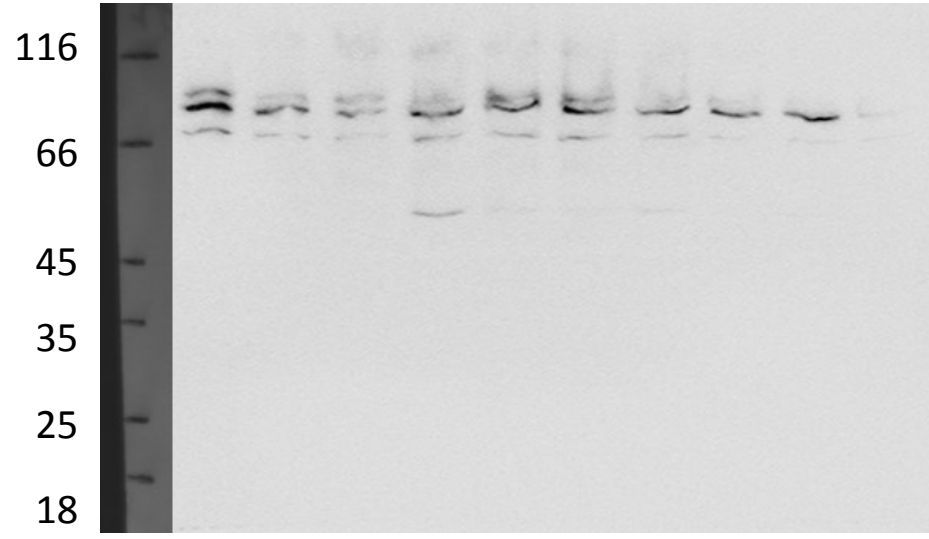
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LCHADD

VLCADD

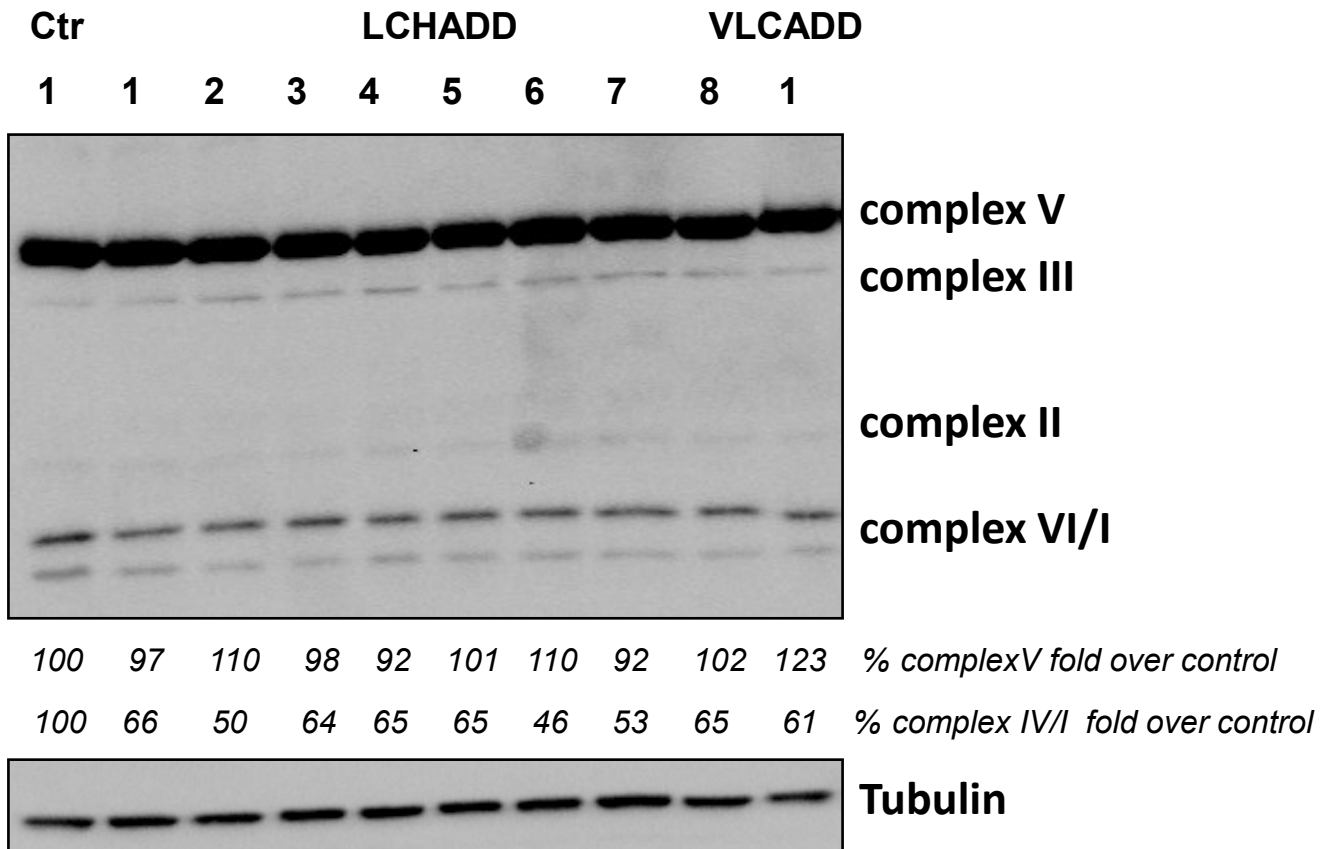
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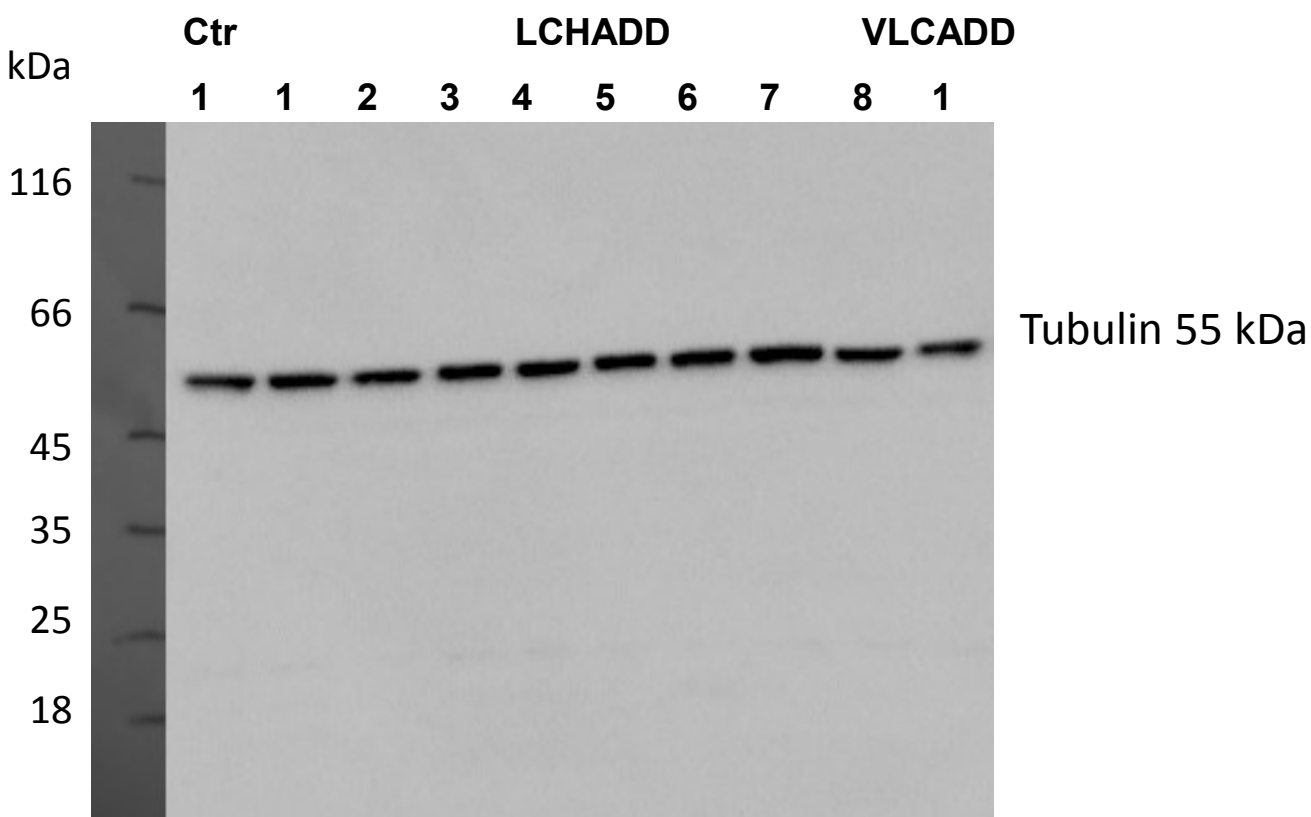
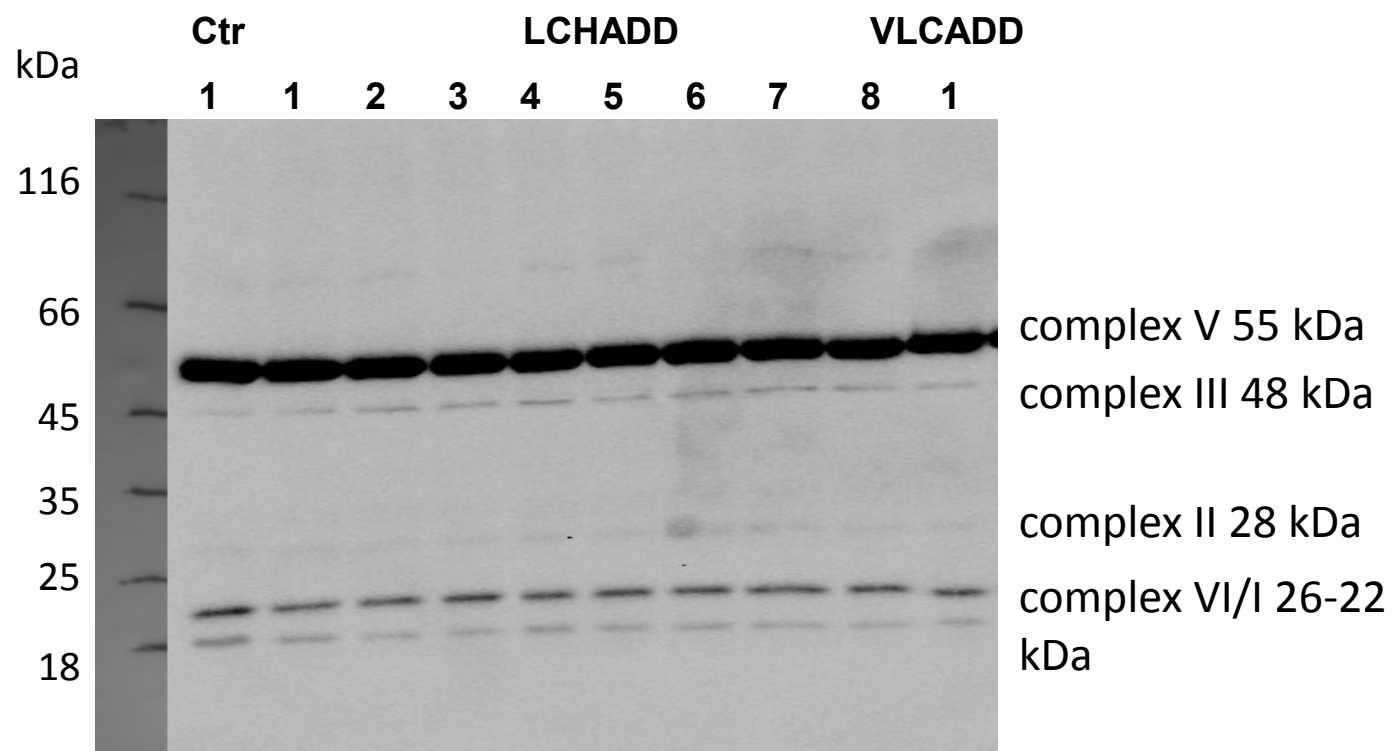
kDa



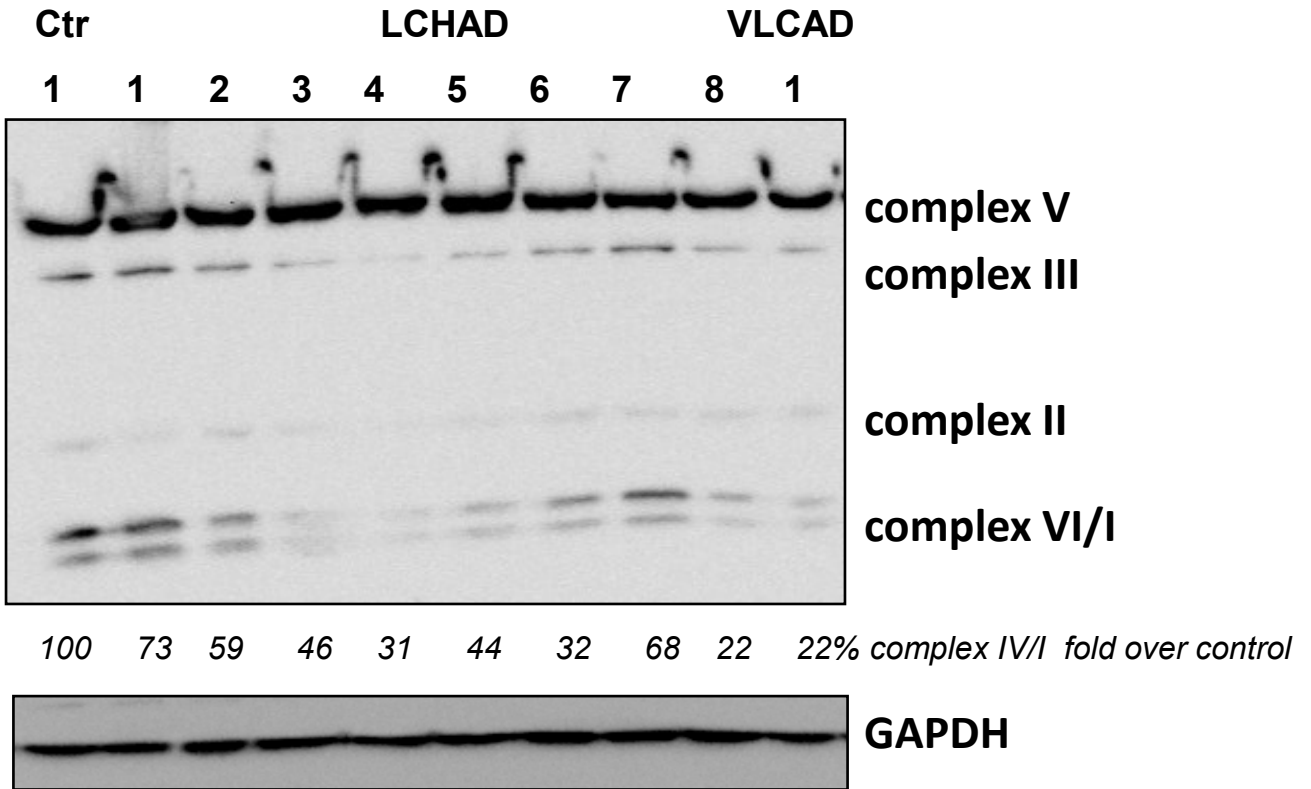
pDNM1LSer673

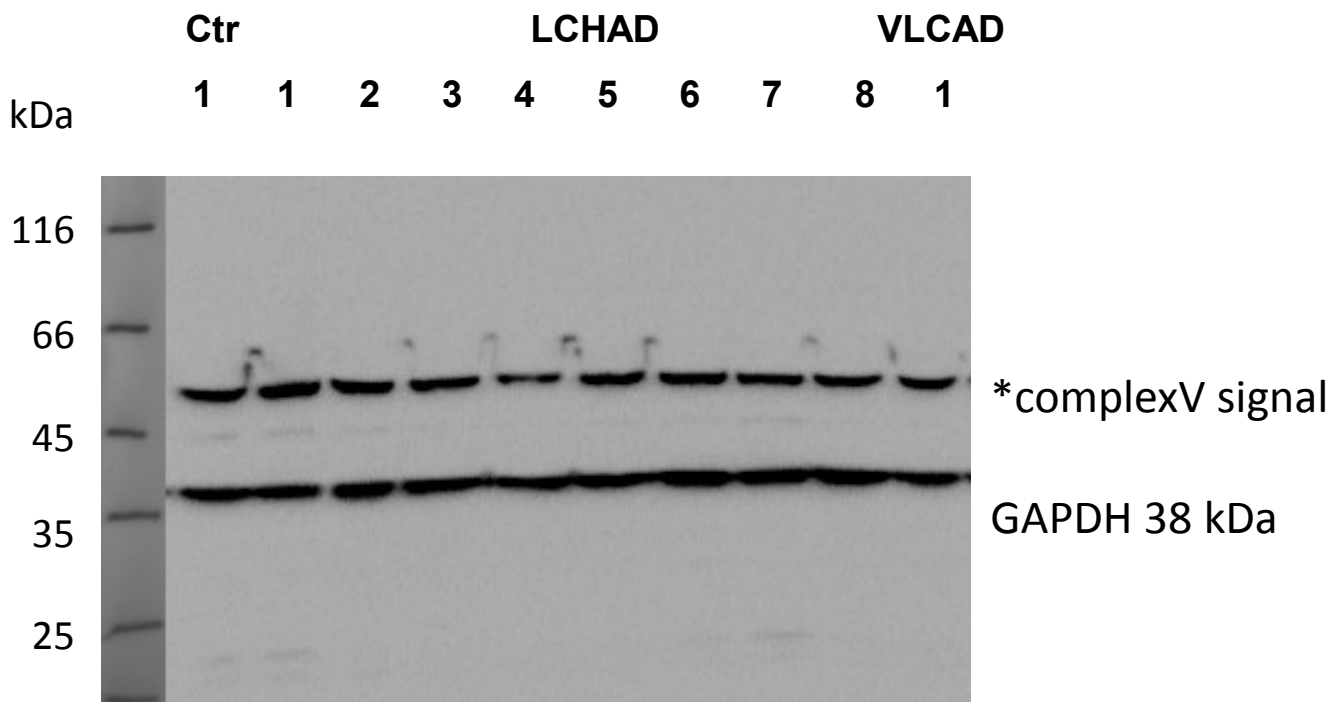
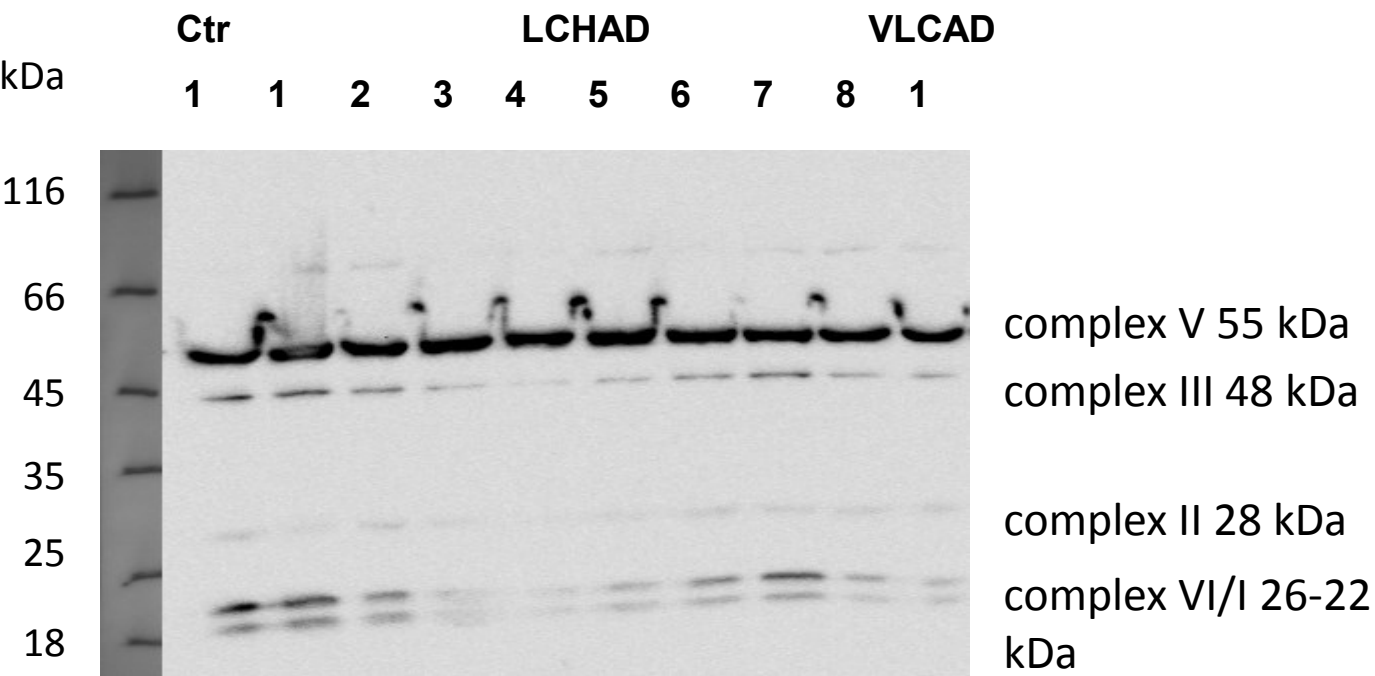
B) Analyses of OXPHOS in LCHADD/VLCADD patients:





Analyses of OXPHOS in LCHADD/VLCADD patients PANEL 2:





Analyses of OXPPOS in LCHADD/VLCADD patients PANEL 3:

