|  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Table e-1: Expression level of CNTN1 in transfected HEK293 cells and cerebellar granule neurons following incubation with patient sera** | | | | | |  | |  | |
| **condition** | **n** | **relative whole cell expression [CNTN1/-actin]** | **normalized whole cell expression [% HC]** | **n** | **relative surface expression [CNTN1/ATPase]** | | **normalized surface expression [% HC]** | |
| **HEK293** |  |  |  |  |  | |  | |
| **untreated** | 8 | 1.06 ± 0.24 | 100 ± 23 | 5 | 0.88 ± 0.07 | | 100 ± 8 | |
| **HC 2d** | 6 | 1.06 ± 0.11 | 100 ± 11 | 7 | 0.64 ± 0.08 | | 100 ± 12 | |
| **HC 4d** | 16 | 0.88 ± 0.07 | 100 ± 8 | 11 | 0.82 ± 0.06 | | 100 ± 8 | |
|  |  |  |  |  |  | |  | |
| **pat1 2d** | 5 | 0.80 ± 0.15 | 75 ± 14 | 7 | 0.40 ± 0.066 | | 66 ± 11 | |
| **pat1 4d** | 6 | 0.90 ± 0.13 | 102 ± 15 | 7 | 0.52 ± 0.075 | | 64 ± 9 | |
| **pat1 2R** | 6 | 0.86 ± 0.22 | 98 ± 25 | 6 | 0.96 ± 0.22 | | 117 ± 27 | |
|  |  |  |  |  |  | |  | |
| **pat3 2d** | 4 | 1.04 ± 0.15 | 98 ± 14 | 5 | 0.60 ± 0.05 | | 99 ± 9 | |
| **pat3 4d** | 6 | 0.80 ± 0.21 | 90 ± 23 | 6 | 0.75 ± 0.2 | | 92 ± 23 | |
| **pat3 2R** | 6 | 0.63 ± 0.12 | 71 ± 13 | 7 | 0.66 ± 0.16 | | 80 ± 19 | |
| **CGN** |  |  |  |  |  | |  | |
| **HC 2d** | 2 | 0.37 ± 0.01 | 100 ± 0.86 |  | n. d. | | n. d. | |
| **HC 4d** | 2 | 0.35 ± 0.05 | 100 ± 5 |  | n. d. | | n. d. | |
|  |  |  |  |  |  | |  | |
| **pat1 2d** | 2 | 0.08 ± 0.01 | 21 ± 3 |  | n. d. | | n. d. | |
| **pat1 4d** | 2 | 0.03 ± 0.002 | 7 ± 0.56 |  | n. d. | | n. d. | |
| **pat1 2R** | 2 | 0.08 ± 0.01 | 20 ± 1 |  | n. d. | | n. d. | |
|  |  |  |  |  |  | |  | |
| **pat3 2d** | 2 | 0.11 ± 0.02 | 29 ± 5 |  | n. d. | | n. d. | |
| **pat3 4d** | 2 | 0.06 ± 0.004 | 18 ± 1 |  | n. d. | | n. d. | |
| **pat3 2R** | 2 | 0.12 ± 0.001 | 39 ± 0.26 |  | n. d. | | n. d. | |

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| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Table e-2: Functional properties of DRG sodium channels incubated with sera of HC or anti-CNTN1 positive patients** | | | | | | | | | | | | | | | |
|  | **HC** | | | | **pat1** | | | | **pat2** | | | | **pat3** | | |
| **condition** | **n** | **cell capacitance (pF)** | **peak current density (pA/pF)** | **n** | | **cell capacitance (pF)** | **peak current density (pA/pF)** | **n** | | **cell capacitance (pF)** | **peak current density (pA/pF)** | **n** | | **cell capacitance (pF)** | **peak current density (pA/pF)** | |
| **1 h** | 23 | 13 ± 0.22 | 292 ± 28 | 8 | | 15 ± 0.64 | 254 ± 28 | 10 | | 15 ± 0.65 | 245 ± 27 | 13 | | 14 ± 0.25 | 230 ± 16 | |
| **2 h** | 18 | 15 ± 0.3 | 270 ± 24 | 11 | | 15 ± 0.42 | 288 ± 18 | 9 | | 14 ± 0.16 | 298 ± 42 | 24 | | 14 ± 0.19 | 316 ± 24 | |
| **24 h** | 25 | 15 ± 0.21 | 247 ± 20 | 15 | | 14 ± 0.36 | 281 ± 38 | 10 | | 16 ± 0.25 | 223 ± 16 | 21 | | 19 ± 0.3 | 212 ± 18 | |
| **48 h** | 26 | 17 ± 0.24 | 280 ± 25 | 19 | | 16 ± 0.22 | 273 ± 22 | 10 | | 14 ± 0.37 | 301 ± 32 | 23 | | 14 ± 0.15 | 341 ± 36 | |
| **72 h** | 32 | 16 ± 0.11 | 261 ± 19 | 12 | | 20 ± 0.47 | 200 ± 23 | 5 | | 14 ± 0.37 | 205 ± 19 | 18 | | 17 ± 0.15 | 185 ± 18 | |

**Figure e-1: Incubation of neurons with anti-CNTN1 positive serum does not lead to changes in expression of pan-neurofascin and βIII tubulin.**

(A-C) Representative images of CGN stained for pan-neurofascin (pan-NF, red) after 3 days of incubation with serum of a healthy control (HC, A) or serum of patient (pat) 1 (B) or pat3 (C). Scale bars: 20 µm. (D-F) Expression of β III tubulin (red) is not altered in CGNs after 3-day incubation with serum of pat1 (E) or pat3 (F) compared to incubation with serum of HC (D). Scale bars: 20 µm. (G-I) Expression of β III tubulin (red) in DRG neurons after 3 days of incubation with serum of pat1 (H) or pat3 (I) compared to the incubation with serum of HC (G). Scale bars: 50 µm. DAPI was used for staining the nucleus (blue).