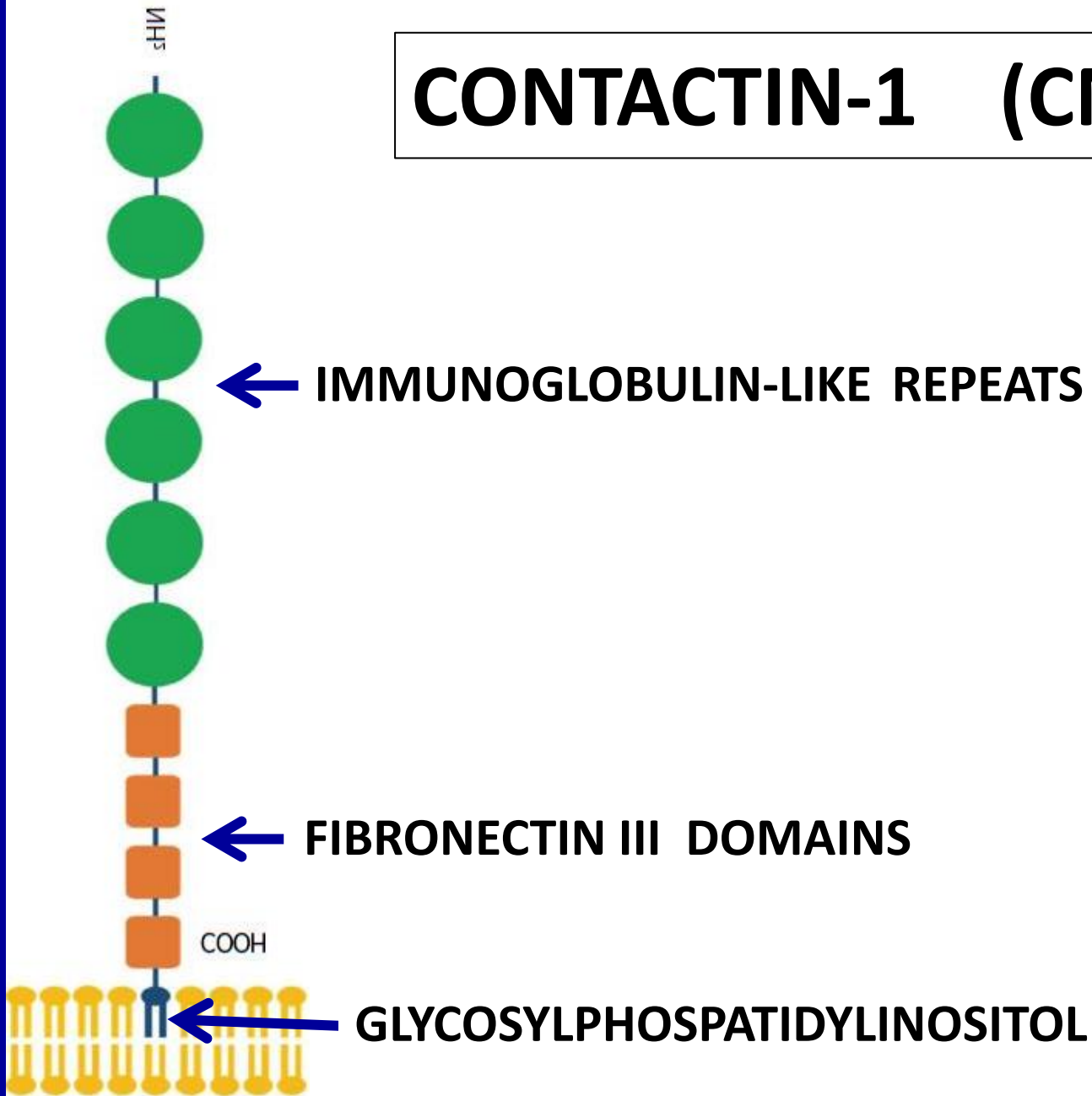


CONTACTIN-1 (CNTN1)



JUXTA PARANODE

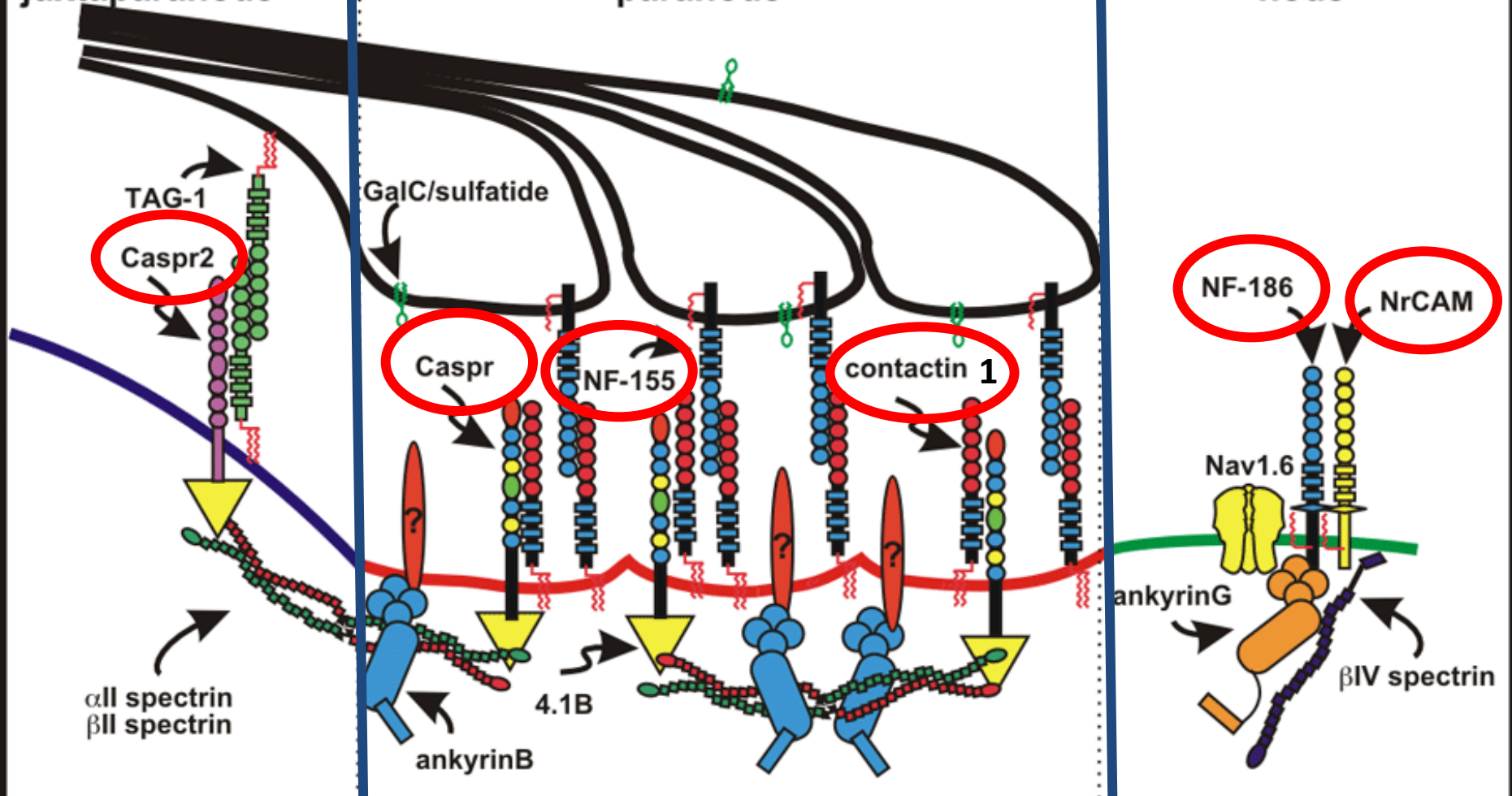
Paranode

Node of
Ranvier

juxtaparanode

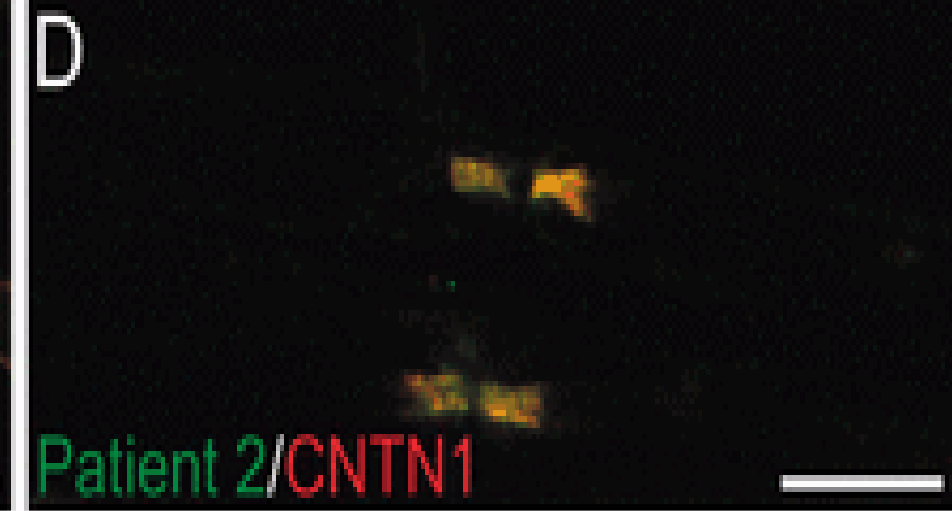
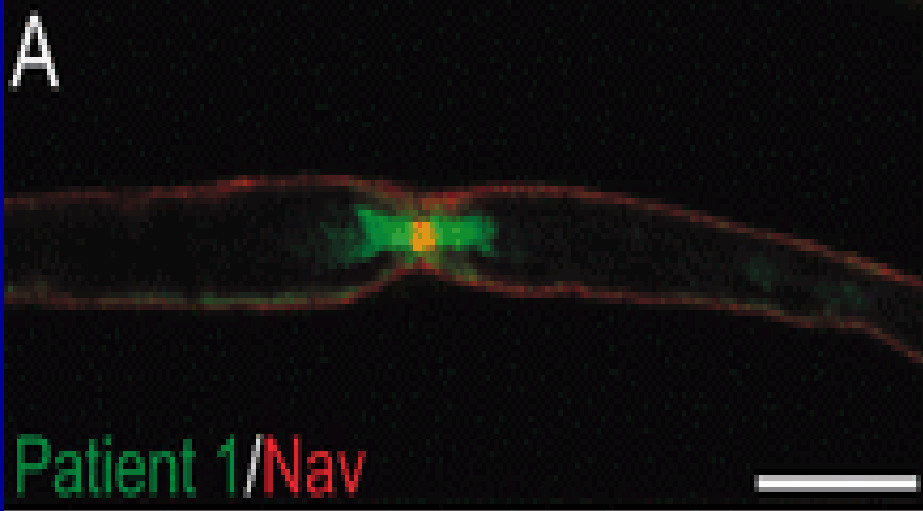
paranode

node



Contactin 1 IgG4 associates to chronic inflammatory demyelinating polyneuropathy with sensory ataxia

Yumako Miura,^{1,*} Jérôme J. Devaux,^{2,*} Yuki Fukami,¹ Constance Manso,² Maya Belghazi,² Anna Hiu Yi Wong,¹ Nobuhiro Yuki^{1,3} and for the CNTNI-CIDP Study Group[†]



Brain. 2015 Jun;138(Pt 6):1484-91.

Contactin 1 IgG4 associates to chronic inflammatory demyelinating polyneuropathy with sensory ataxia.

Miura Y¹, Devaux JJ², Fukami Y¹, Manso C³, Belghazi M³, Wong AH¹, Yuki N⁴; CNTN1-CIDP Study Group.

J Neurol Neurosurg Psychiatry. 2015 Jul;86(7):720-8.

Destruction of paranodal architecture in inflammatory neuropathy with anti-contactin-1 autoantibodies.

Doppler K¹, Appeltshauer L¹, Wilhelmi K¹, Villmann C², Dib-Hajj SD³, Waxman SG³, Mäurer M⁴, Weishaupt A¹, Sommer C¹.

Brain. 2016 Mar 26. pii: aww062.

Contactin-1 IgG4 antibodies cause paranode dismantling and conduction defects.

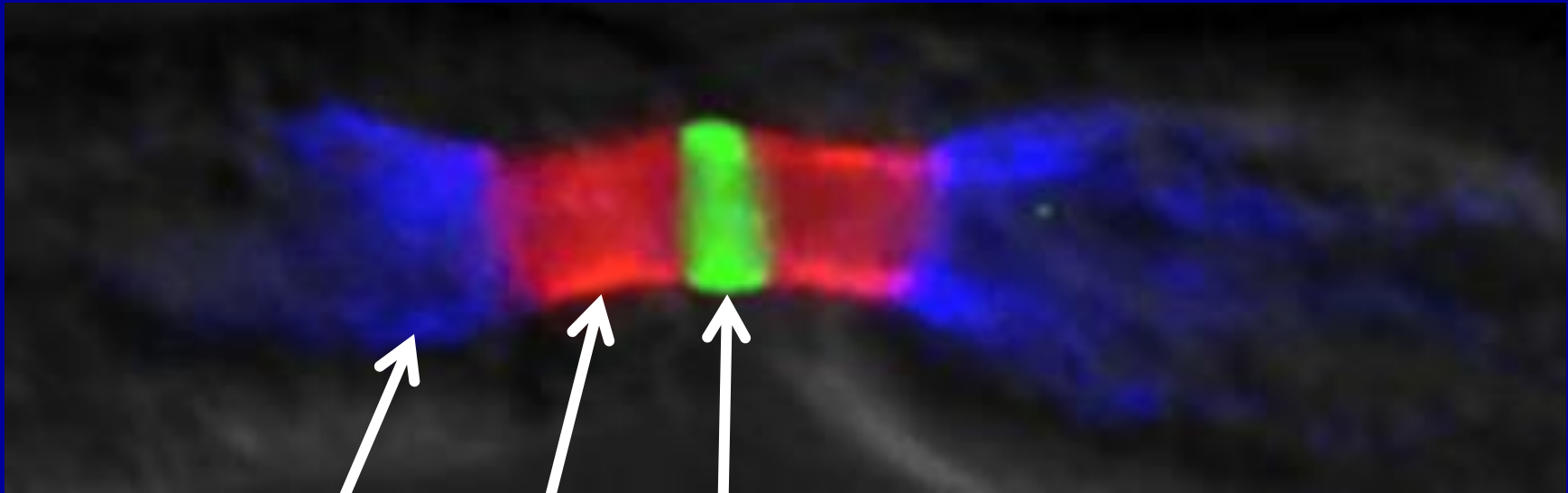
Manso C¹, Querol L², Mekaouche M¹, Illa I², Devaux JJ³.

Rituximab in treatment-resistant CIDP with antibodies against paranodal proteins

Luis Querol, MD, PhD, Ricard Rojas-García, MD, PhD, Jordi Diaz-Manera, MD, PhD, Joseba Barcena, MD, Julio Pardo, MD, Angel Ortega-Moreno, MD, Maria Jose Sedano, MD, Laia Seró-Ballesteros, MD, Alejandra Carvajal, MD, Nicolau Ortiz, MD, PhD, Eduard Gallardo, PhD and Isabel Illa, MD, PhD

NODO - PARANODOPATHY

AUTOANTIBODIES



NODE

*Neurofascin 186 ,
Glyomedin , NrCam*

PARANODE

*Neurofascin 155 , Contactin 1
Caspr 1*

JUXTAPARANODE

Caspr2 , Contactin 2

NODO - PARANODOPATHY

SEVERE POLYRADICULONEUROPATHY

GBS - LIKE

SENSORIMOTOR NEUROPATHY

PREDOMINANT DISTAL WEAKNESS

CHRONIC COURSE

ATAXIA

SEVERE TREMOR

POOR RESPONSE TO IVIg

ANTIBODIES TO NEUROFASCIN 155

4 / 117	J.King	2012
8 / 254	J.King	2012
4 / 16	JJ.Devaux	2012
0 / 16	N.Kawamura	2013
2 / 53	L.Querol	2014
13 / 533	Y.Miura	2014
4 / 53	K.Doppler	2015
9 / 43	H.Ogata	2015 (IgG4)

ANTIBODIES TO CONTACTIN 1

1 / 50	JJ.Devaux	2012
4 / 46	L.Querol	2013
13 / 533	Y.Miura	2014 (IgG4)
4 / 53	K.Doppler	2015

ANTIBODIES TO NODAL PROTEINS

%	NF 186	GLYOMEDIN	NcCAM	CONTACTIN 1
GBS	16	12	4	12
AIDP	12	14	6	16
AMAN	18	10	2	8
CIDP	12	6	2	16

J.J. Devaux, M.Okada, N.Yuki 2012

J Neuroimmunol. 2014 Nov 15;276(1-2):207-12.

Autoantibodies to neurofascin-186 and gliomedin in multifocal motor neuropathy.

Notturmo F¹, Di Febo T², Yuki N³, Fernandez Rodriguez BM⁴, Corti D⁴, Nobile-Orazio E⁵, Carpo M⁶, De Lauretis A⁷, Uncini A⁸.

**MMN n= 53 : 62% (IgG)
: 12% (IgM)**

PLoS One. 2015 Jul 28;10(7)

Contactin-1 and Neurofascin-155/-186 Are Not Targets of Auto-Antibodies in Multifocal Motor Neuropathy.

Doppler K¹, Appeltshauser L¹, Krämer HH², Ng JK³, Meinl E³, Villmann C⁴, Brophy P⁵, Dib-Hajj SD⁶, Waxman SG⁶, Weishaupt A¹, Sommer C¹.

**MMN n= 33 : 0 (IgG)
: 0 (IgM)**