



400

2017

RelA

せ
に

4

RelA

【 っ ⁵

| っ 。そっ 、 せ 【
 っ 、 せ TLR ⁶
 TLR TLR7/8/9 っ 。そっ
 IRF7 ⁷ MyD88 | っ 。そっ
 | っ 。そっ 2011 Crow
 | っ 。そっ
 Aicardi-Goutières せ に せ
⁸ ず
 ず、 | っ 。そっ

せ

せ

そ 。す

RELA

5

6

RelA

っ

RelA

っ

RelA っ

| IFN こ ⁹

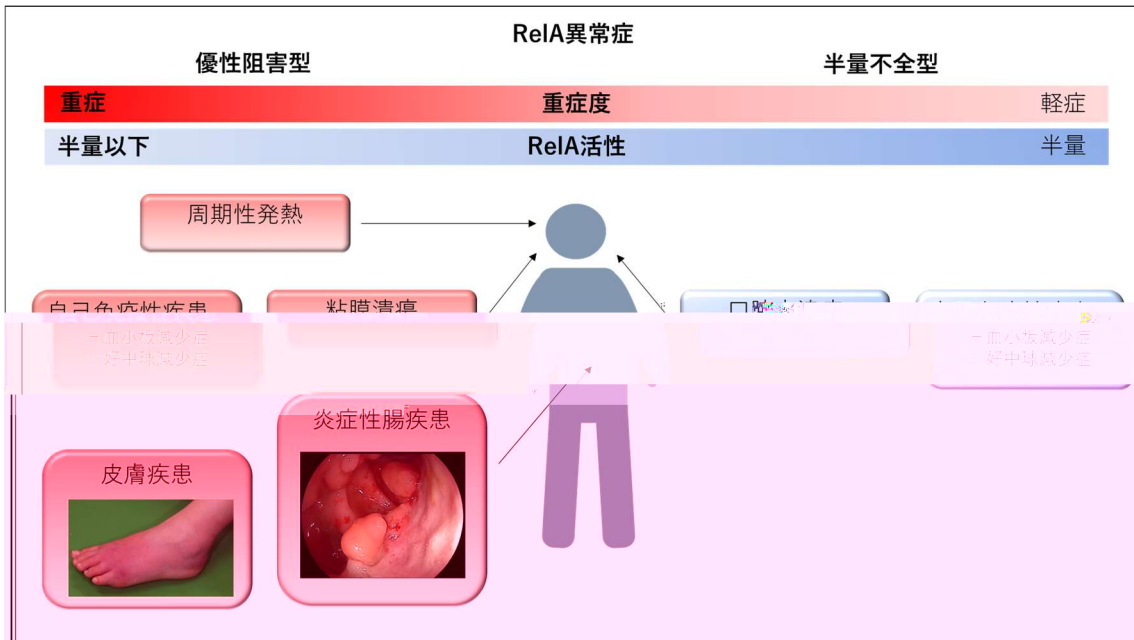
TLR7 | っ 。そっ っ 。そっ INF- α 2, INF- β , INF- γ 1, INF- γ 2/3

っ 10 っ せ せ RNA mDC

pDC TLR7
 ずっ IRF7 MyD88
 IRF7 | っ 。そっ
 NF κ B RelA っ TLR7 IRF7
 | っ 。そっ

NF κ B | っ 。そっ RelA

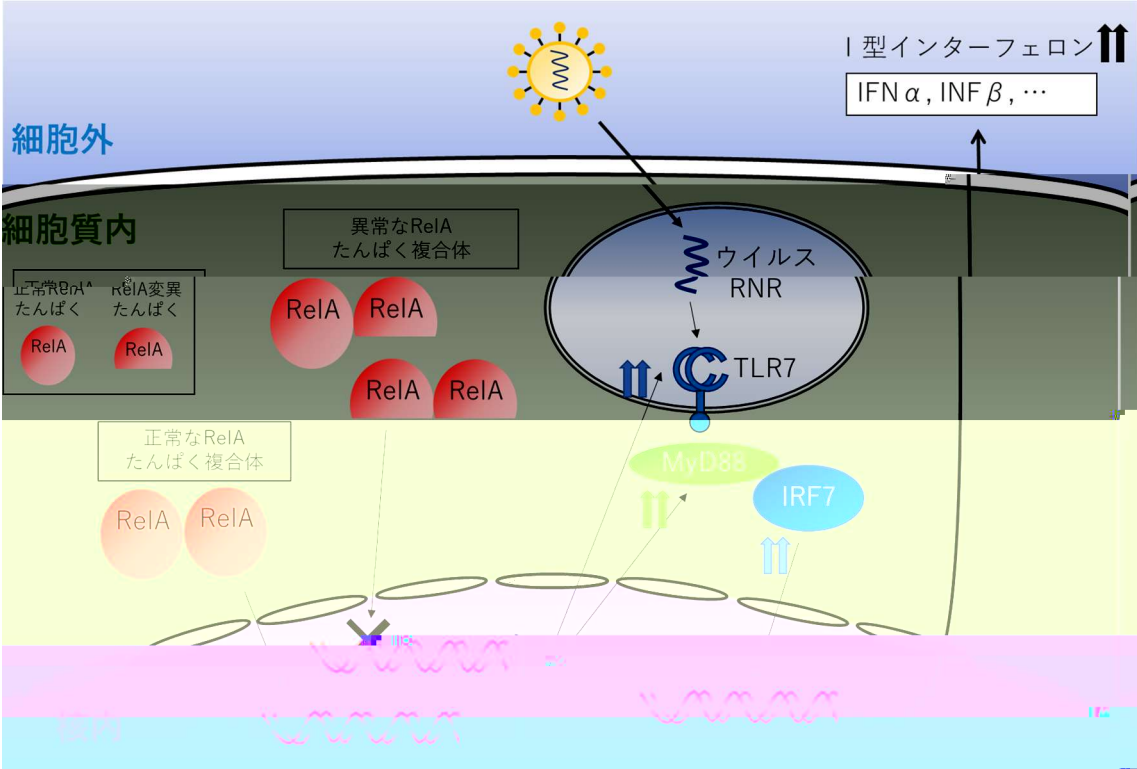
| っ 。そっ



1

RelA

RelA



2. RelA

RelA つ

RelA つ 。

RelA つ 。

TLR7 MyD88

IFNα INFβ

JP21K07791

AMED

1. RELA

RelA つ

NF B せ

RelA つ DNA

NF B

2. 1

3. | つ 。 そつ IFN- α IFN- β 、 せ
4. 1
5. 【 つ
6. TLR:Toll Toll-like receptor
7. IRF7: つ 。 そつ 7 interferon regulatory factor 7
| つ 。 そつ
8. Aicardi-Goutières TREX1,IFIH1
| つ 。 そつ
9. IFN こ | つ 。 そつ
10. つ せ せ RNA つ 1

せ Human RELA dominant-negative mutations underlie type I interferonopathy with autoinflammation and autoimmunity

Kunihiko Moriya ^{§ *}, Tomohiro Nakano [§], Yoshitaka Honda, Miyuki Tsumura, Masato Ogishi, Motoshi Sonoda, Masahiko Isa-Nishitani, Takashi Uchida, Mohamed Hbibi, Yoko Mizoguchi, Masataka Ishimura, Kazushi Izawa, Takaki Asano, Fumihiko Kakuta, Daiki Abukawa, Darawan Rinchai, Peng Zhang, Naotomo Kambe, Aziz Bousfiha, Takahiro Yasumi, Bertrand Boisson, Anne Puel, Jean-Laurent Casanova, Ryuta Nishikomori, Shouichi Ohga, Satoshi Okada*, Yoji Sasahara, Shigeo Kure ^{§ *}

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