Haploinsufficiency of A20
- Heterozygous mutations, LOF
  - Defective K63 DUB activity
  - Increased NF-κB activity, Increased NLRP3 inflammasome activity
  - Targeted cytokine therapies

Otulipenia
- Bi-allelic mutations, LOF
  - Defective linear DUB activity
  - Increased NF-κB activity
  - Targeted cytokine therapies

Target genes:
- Proinflammatory cytokines

Nucleus

Figure 1