Suppl. Table S2: Review of the literature regarding eculizumab treatment in patients with SVs in the CFH gene cluster. The term “n/a” was used as representative of “not mentioned / information not available”. \*Breville et al. “Eculizumab was considered but not retained due to stabilization of laboratory parameters and a joint decision between the medical staff and the patient after presenting the risks, benefits, and economic burden. Eculizumab was retained as a second line treatment option in case of nonresponse or premature relapse”.50 †Togarsimalemath et al. “[Eculizumab] was not available in the family’s country”.51 ‡Tortajada et al. Patients were treated with angiotensin converting enzyme inhibitor therapy. Eculizumab was mentioned as potential beneficial therapy but was not mentioned as treatment for the patients in this study.53 §Gale et al. Eculizumab is mentioned as potential beneficial treatment but it was not mentioned that eculizumab was given to the patients in this study.54

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
| **Structural variant** | **Disease** | **Eculizumab therapy** | **outcome** | **Reference** |
| *CFH/CFHR1* hybrid gene | aHUS, | yes | beneficial | 44, 32 |
| *CFH/CFHR3* hybrid gene | aHUS | yes | beneficial | 9 |
| *CFHR3/CFHR1* deletion | aHUS,  DEAP-HUS | yes | beneficial | 46 |
| *CFHR3/CFHR1* deletion | SLE, TMA | yes | beneficial | 47, 48 |
| *CFHR4/CFHR1* deletion | aHUS, DEAP-HUS | n/a | n/a | 49, 22 |
| *CFHR4/CFHR1* deletion | IgG4-RD | no\* | n/a | 50 |
| *CFHR3/CFHR1* hybrid gene | C3G | n/a | n/a | 14 |
| *CFHR1/CFH* hybrid gene | aHUS | yes | beneficial | 7 |
| *CFHR1/CFHR5* hybrid gene | C3G | no† | n/a | 51 |
| *CFHR2/CFHR5* hybrid gene | C3G-DDD | yes | no effect | 15 |
| *CFHR5/CFHR2* hybrid gene | C3G | n/a | n/a | 52 |
| Intra *CFHR1* duplication | C3G | no‡ | n/a | 53 |
| Intra *CFHR5* duplication | C3G | no§ | n/a | 54 |
| Intra *CFHR5* duplication | C3G | n/a | n/a | 55 |
| Intra *CFHR3* deletion | C3G | n/a | n/a | 13 |
| *CFHR3/CFHR4* hybrid gene | C3G | n/a | n/a | 13 |
| *CFH* deletion | IC-MPGN | n/a | n/a | 13 |
| *CFHR4/CFHR1* duplication | C3G, IC-MPGN | n/a | n/a | 13 |