

Table 5. Comparison of Clinical Characteristics, Genotype, and Allele Frequency of the Prevalent Variants between Childhood-onset and Adult-onset Stargardt Disease with two or more disease-causing *ABCA4* variants.

	Childhood-onset Stargardt disease (n=34)						Adult-onset Stargardt disease (n=64)						
Median age of onset (yrs)	8.5 (3-14)						27.0 (17-65)						
Median age at examination (yrs)	11.5 (7-16)						44.0 (21-71)						
Median LogMAR VA of the right eye	0.79 (0.18-1.30)						1.00 (-0.08-2.00)						
	Total (n=32)						Total (n=64)						
	Grade 1 1	Grade 2 1	Grade 3a 8	Grade 3b 22	Grade 3c 0	Grade 4 0	Grade 1 0	Grade 2 6	Grade 3a 4	Grade 3b 34	Grade 3c 12	Grade 4 8	
Fundus Appearance	Macular 2		Flecks Peripheral 21		No flecks 9		Macular 14		Flecks Peripheral 39		No flecks 11		
			Pigmentation 2		No pigmentation 30				Pigmentation 29		No pigmentation 35		
Autofluorescence Pattern	Type 1 8		Total (n=29) Type 2 21			Type 3 0		Type 1 20		Total (n=62) Type 2 33		Type 3 9	
OCT, CFT (µm) of the right eye	Total (n=19) 61.0 (33-138)						Total (n=33) 81.0 (20-297)						
ERG group	Group 1 7		Total (n=18) Group 2 1		Group 3 10		Group 1 34		Total (n=59) Group 2 7		Group 3 18		
Genotype group classification	Group A 7		Total (n=34) Group B 15			Group C 12		Group A 3		Total (n=64) Group B 26		Group C 35	
Frequencies of the most prevalent variants	c.5461-10T>C 8 (11.8%)		p.Gly1961Glu 4 (5.9%)			p.Cys1490Tyr 4 (5.9%)		p.Gly1961Glu 16 (12.5%)		p.Gly863Ala 13 (10.1%)		p.Leu2027Phe 8 (6.3%)	

AF type = autofluorescence type; CFT = central foveal thickness; ERG = electroretinogram; LogMAR VA = logarithm of the minimum angle of resolution visual acuity; OCT = optical coherence tomography.

In order to investigate the differences between the patients with childhood-onset Stargardt Disease (STGD) and those with adult-onset STGD, clinical and molecular genetic data of patients with adult-onset STGD ascertained at Moorfields Eye Hospital were reviewed. The comparison group consisted of all patients who had adult-onset STGD (older than 17 years old), and two or more disease-causing *ABCA4* variants. For the purpose of this comparison, 34 patients with childhood-onset STGD and two or more disease-causing *ABCA4* variants were selected.