We consider the problem of estimating the lifetime-risk of a disease with variable age at onset correcting for ascertainment. In addition to the proband, our working sample includes all first-degree relatives (sibs and/or parents) of the proband. We show that the likelihood corrected for ascertainment is equal to the likelihood without any reference to family membership times a correction term. We propose formulae for the correction term in the case of complete ascertainment, or sampling families with at least one diseased individual in the family with equal probability. We present results from a simulation study that demonstrates the validity and performance of the methodology proposed. We extend the theory to obtain estimates of penetrance for rare susceptibility genes for diseases with variable age at onset. As an example, we present estimates of penetrance for BRCA1 gene from a population-based family study, where we show that the risk of breast cancer among carriers by age 55 is 0.28 [95%CI 0.10-0.36].