Author Response: Prevalence of Corneal Dystrophies in the United States: Estimates from Claims Data

Møller and colleagues[1] raise a cogent observation in noting that relying on claims data to reveal information on prevalence of corneal dystrophies has important limitations, especially regarding diagnostic accuracy. While we may not have discussed this limitation as fully as the letter's authors would have desired, we do identify the source of these diagnostic codes—"an eye care provider (ophthalmologist or optometrist)"—and we point out in our "Discussion" that coding, "particularly with endothelial dystrophy," is subject to inaccuracies in claims data.[2] It goes without saying that a diagnostic code for a specific corneal dystrophy in claims data does not provide assurance that the diagnosis was verified by electron microscopy, histology, family history, or genetic tests.

We contend that including information on dystrophies that were assigned an ICD-9-CM code of "endothelial dystrophy" should be included. Truly, they represent what the authors term "an etiologically heterogenous group of diseases of a degenerative nature" and we share the authors' anticipation of better delineation of corneal dystrophies that will come from applying the new IC3D coding. However, equally truly, a genetic basis for endothelial dystrophies has been vetted by Baratz et al.,[3] and endothelial dystrophies represent a major cause of corneal transplantation in the United States. Such surgery is typically performed by highly trained corneal specialists whose diagnostic coding is based on a thorough evaluation of the cornea and is likely to be accurate.

Indeed, the information we provided does estimate prevalence rather than frequency, as it fits the epidemiologic definition of prevalence (the number of cases in a given population at a designated time). Studies that rely on claims data surely are subject to limitations, and associations uncovered in such studies should be viewed as hypothesis-generating rather than causal in nature. They also provide important insights into the health care burden imposed by corneal dystrophies, as claims data reflect actual care provision for the specified condition, which can lead to better estimates of cost.

We look forward to future reports that estimate the prevalence of corneal dystrophies in defined populations and rely on more accurate coding of corneal dystrophies, which—as Møller et al. note—is needed.

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