Table 2: Published X-linked, *COL4A5* ATS mutations, characterized by absent or very late onset ESKD, absent ocular complications and absent or delayed neurosensory deafness. Ultrastructurally these patients show intermediate GBM changes and mostly present as phenocopies of TBMN with microscopic hematuria.

X-linked COL4A5 Mutations	Clinical findings and patients' characteristics	References
C1564S L1649R R1677Q	The three most common "benign" "adult type" X-linked *COL4A5* mutations in Utah/USA. Delayed or absent ESKD with only late neurosensory deafness	Barker et al, 1996[7] Pont-Kingdon et al, 2009[6] Martin et al 1998[10]
G624D P628L	Benign familial hematuria & diffuse thinning of the GBM. Benign clinical course with absence of or late ESKD Absent ocular complications and no neurosensory deafness	Martin et al, 1998[10] Slajpah et al, 2007[11] Demosthenous et al, 2012[13]
G156A	A Chinese family with TBMN and only microscopic hematuria	Chen et al, 2001[8]
C1638Y	A large family in New Zealand. Only 3 out of 8 males progressed to ESKD. No ocular problems or deafness	Wilson et al, 2007[12]
G1000V	A Japanese family with only benign familial hematuria	Kaneko et al, 2010[9]