Records identified through database searching (PubMed / Web of Science) (n = 1006)

Additional records identified through other sources (n = 15)

Records after duplicates removed (n = 605)

Records screened (n = 605)

Records excluded for review:
- non-English (n = 70)
- not relevant (n = 35)
- short conference proceeding or meeting abstract (n = 49)

Full-text articles assessed for eligibility (n = 451) (included in review)

Studies included in qualitative synthesis (n = 53)

Studies included in quantitative synthesis (n = 46)

Full-text articles excluded for meta-analysis:
- not mentioning auditory phenotype and genotype (n = 398)

Full-text articles excluded for quantitative analysis:
- patients described in another included paper (n = 5)
- patients with a second genetic disorder (n = 1)
- unclear phenotype (n = 1)