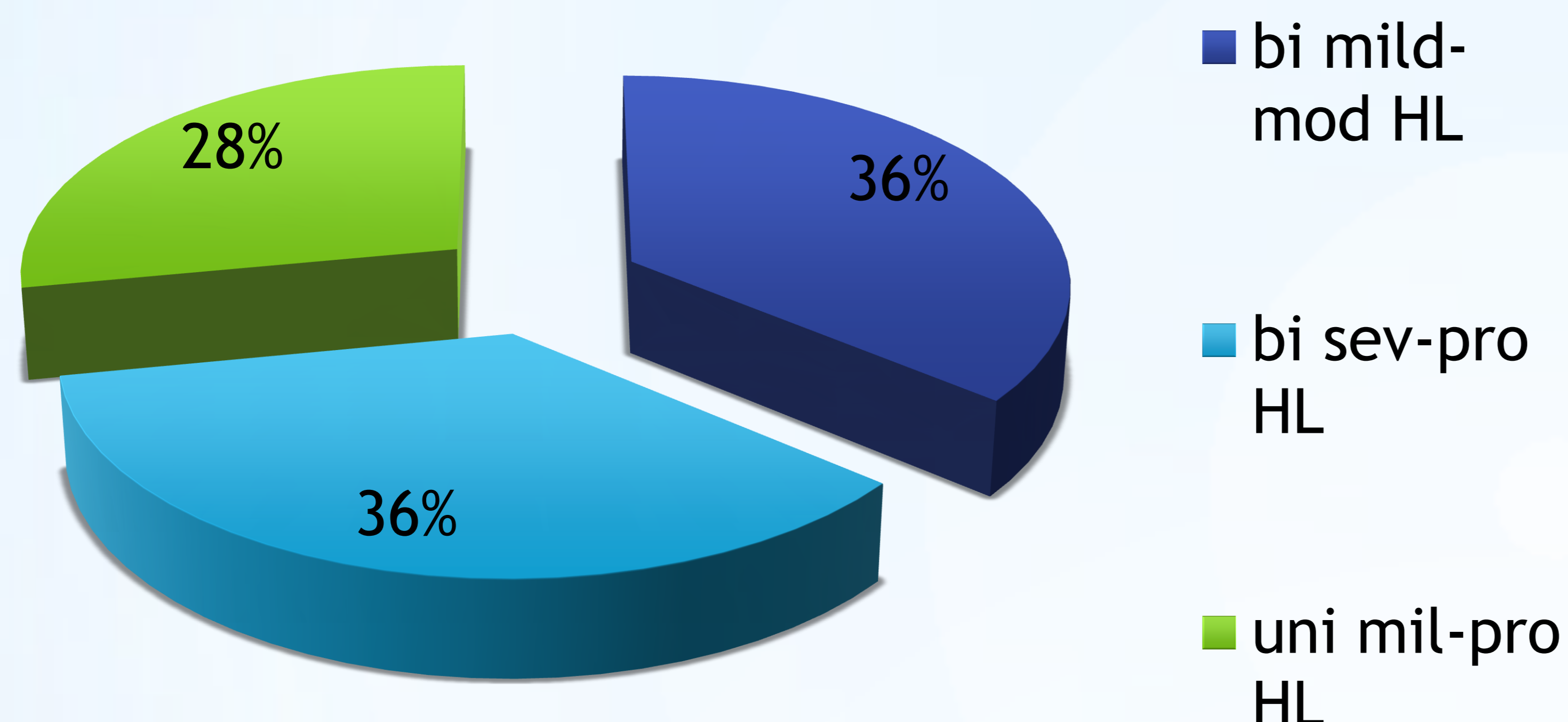


The aetiology yield in Permanent Childhood Hearing Impairment (PCHI) from implementing the BAAP guidelines

AIMS

To measure the outcomes of offering structured aetiological investigation of PCHI in children in line with recommendations from the BAAP/BAPA Guidelines.

Population investigated



METHOD

We retrospectively applied standards, set by the British Association of Audiovestibular Physicians on the aetiological investigation of children with permanent hearing impairment¹.

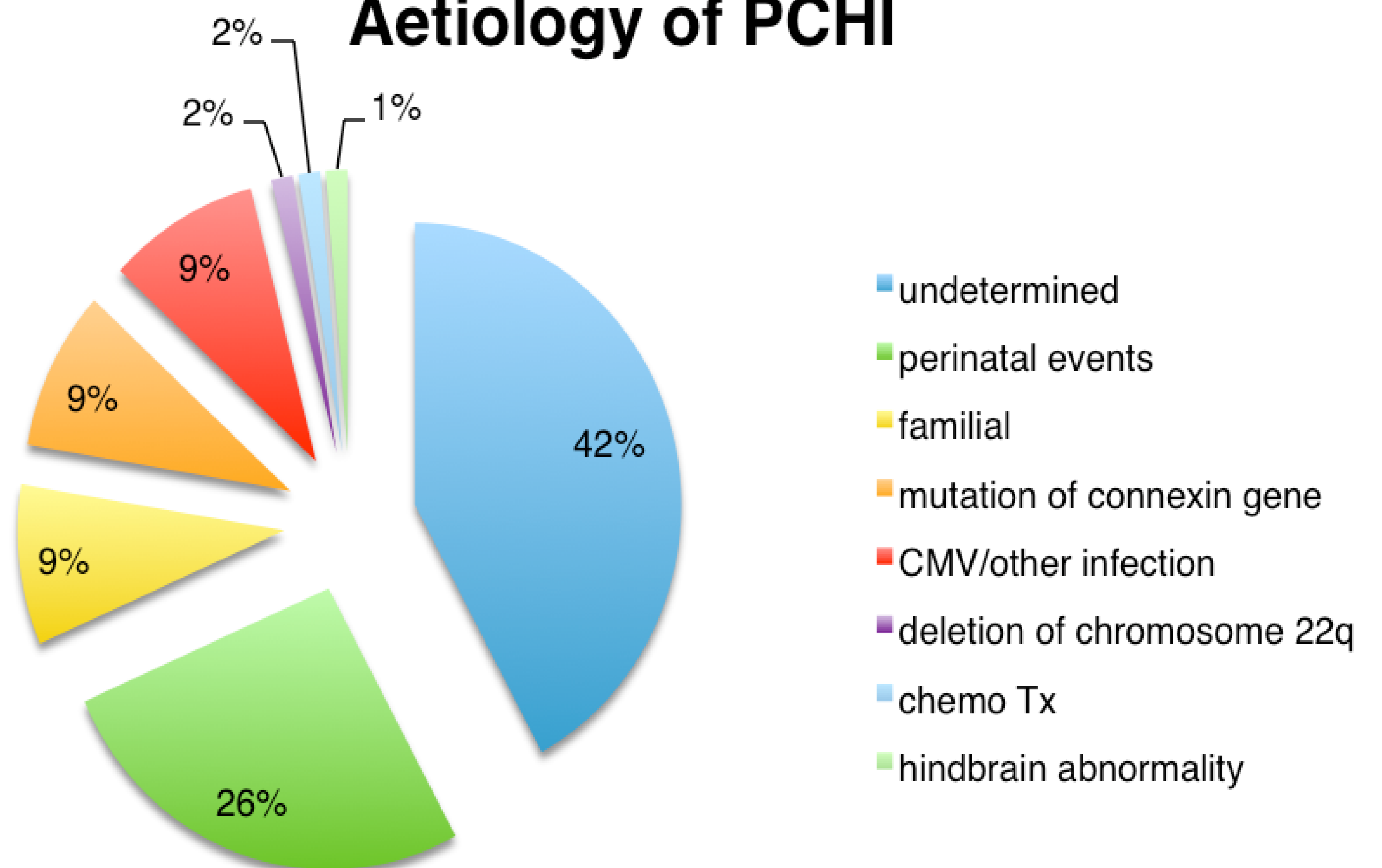
Children aged 0-19 were identified from the Audiology department database of a Foundation Trust serving a population of 0.5 million. 152(92%) were invited to attend for assessment, 89(54%) responded and 76(46%) agreed to proceed.

The medical records of non-responders were reviewed and 21(26%) had already been investigated or had an established diagnosis/syndrome known to be associated with PHI. All responders were investigated to BAAP standards.

RESULTS

- 27(36%) had bilateral mild to moderate HL, 27(36%) severe-profound HL, and 22 (28%) unilateral mild - severe .
- In 32(42%) children the aetiology was undetermined or investigations were on-going at the time of writing.
- PCHI was attributable to perinatal events in 20(26%) children.
- In 7(9%) children PCHI was familial and in a further 7(9%) a mutation of the connexin gene was found.
- CMV or other infections accounted for 7(9%) and among the remaining children 1 had a deletion of chromosome 22q, 1 PCHI was secondary to chemotherapy and 1 child had a hind-brain abnormality

Aetiology of PCHI



CONCLUSION

The structured aetiological investigation of PCHI in children has many benefits not least to answer parent's question "why is my child deaf".

In addition we have identified previously unsuspected diagnoses such as Connexin 26 mutations and 22q deletion, which is important as these children and their families can receive genetic counselling, and appropriate onward referral

We have also developed local pathways following diagnosis of PCHI.