Investigating the occurrence of Charles Bonnet syndrome (CBS) features in patients diagnosed with inherited retinal disease (IRD)

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Background

- In the UK, inherited retinal diseases (IRDs) are reported to be the leading contributor of certifiable blindness in the working age population (20.2%). (1)
- Treatment options are limited despite advances in gene therapy and new symptoms such as visual hallucinations are also of concern. (2,3)
- The prevalence of Charles Bonnet syndrome in people with poor vision is uncertain, with estimates suggesting 20% of patients may be affected. (4)
- The clinical course of IRDs often involves severe and progressive vision loss, presenting a risk for developing CBS (6), but likely underexplored.



Figure 1. A selection of patients' drawings of visual hallucinations experienced in CBS.(5)

Purpose

To identify whether there is an unmet need within the inherited retinal disease patient population in regards to addressing potential Charles Bonnet syndrome symptoms.

Methods

- This project was approved by the hospital Audit and Clinical Effectiveness department.
- A bespoke questionnaire with multiple choice answers and space for free text was designed following a detailed literature review.
- Patients > 18 years old with a diagnosis of IRD under St Paul's Eye Unit were randomly selected from an electronic database and contacted by telephone, and invited to complete the questionnaire, either immediately following the initial contact or at a later convenient time and date, over the telephone.
- Additionally, all patients attending the dedicated IRD clinic were also invited to complete a paper questionnaire at their appointment.

Results

- 50 questionnaires were completed between 26/05/2022 and 16/06/2022, of which 44 were suitable for analysis (excluding incomplete questionnaires).
- Twenty-seven respondents were female, rest male. Mean age of all respondents was 53.66 years ± 16.80 years.
- Eight respondents (18.18%) reported experiencing visual hallucinations, 2 were unsure and 34 reported experiencing no visual hallucinations.
- Mean visual acuity was lower in patients reporting visual hallucinations (LogMAR 1.20 ± 0.95) than those that did not (LogMAR 0.64 ± 0.78).
- Respondents with IRDs typically affecting the macula had a higher incidence of visual hallucinations (1 in 5) when compared to IRD associated with preserved central vision (1 in 9).
- The majority of respondents had no prior knowledge of CBS (77.27%) and did not want/ have a preference on being informed about CBS at the time of diagnosis of IRD (79.55%).
- Of the 8 reporting visual hallucinations, only 2 felt this affected their life negatively, with 6 reporting it had no effect.
- Mean index of multiple deprivation was 3.95 ± 3.14 and was highest in respondents who reported experiencing visual hallucinations (2.38 ± 2.96).

Conclusions

- There may be a need to question IRD patients directly about visual hallucinations due to an apparent prevalence (18%) within this cohort and poor awareness of CBS.
- Only a minority of patients with self-reported visual hallucinations wished for additional support; patients may develop coping mechanisms and habituation without clinical input.
- Screening questions may be required for select IRD patients (e.g. macular dystrophies) to ensure CBS features are managed in line with the patient's preferences.

Figures

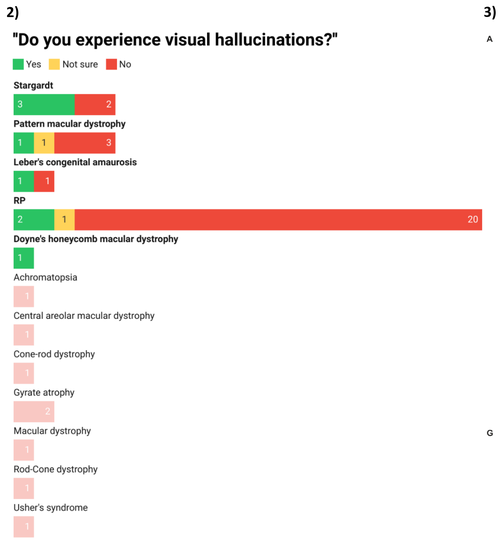


Figure 2. Bar chart demonstrating total number of IRDs included from completed questionnaires and proportion of respondents experiencing visual hallucinations. (Retinitis pigmentosa = RP).

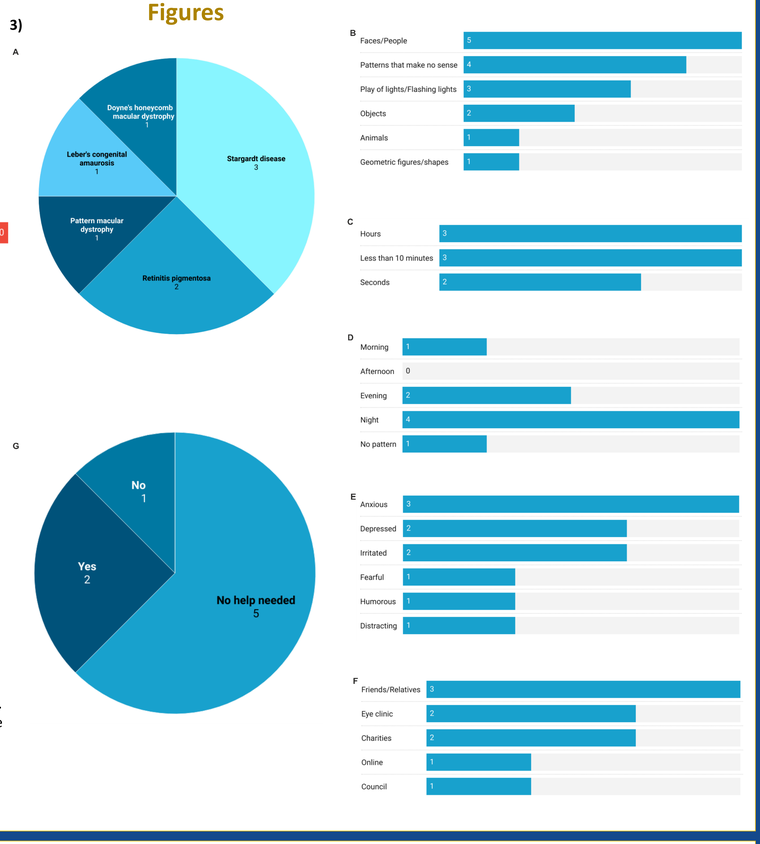


Figure 3. A) Pie chart demonstrating the proportion of IRD diagnoses in respondents who experienced visual hallucinations. B) Bar chart demonstrating the reported subjects of hallucinations. C) Bar chart demonstrating reported typical duration of visual hallucinations per week. D) Bar chart demonstrating the typical timings of hallucinations during the day for individuals. E) Bar chart demonstrating the effect visual hallucinations had on the mood of the individual. F) Bar chart demonstrating sources of help accessed for symptoms of visual hallucinations by individuals. G) Pie chart demonstrating perceptions of adequate levels of support for visual hallucinations

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