Emotion Recognition in People with Huntington's Disease: A Comprehensive Systematic Review

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Abstract

Background: Deficits of emotion recognition have received increasing attention in people with Huntington's disease (HD) in the three decades since the discovery of the HD gene. However, the characterisation of such deficits across different disease stages, types of stimuli, and sensory modalities is currently unclear.

Objective: This study aimed to provide a comprehensive review of the evidence on emotion recognition deficits in HD gene carriers (both manifest and premanifest) over the three decades since definitive gene testing.

Method: A systematic review was carried out from January 1993 to January 2025 across MEDLINE, PsycINFO, Academic Search Complete, and CINAHL (PROSPERO registration: CRD42023398649).

Results: From 9735 initial citations, 59 studies were eventually included. In manifest HD, facial recognition of negative emotions such as anger, fear, disgust, and sadness was consistently impaired, whereas happiness and neutral expressions were generally spared. A few auditory studies showed consistent deficits for disgust, fear, and anger, while happiness and sadness appeared less affected. Only preliminary evidence is currently available for deficits involving body language, visual and written vignettes, videos, and olfactory and gustatory tasks. Although sparser, the evidence for premanifest HD suggests that some individuals may develop significant recognition difficulties prior to motor onset, particularly due to early frontostriatal deterioration and white matter disruption.

Conclusions: Impairments of facial recognition of negative emotions are reported consistently in manifest HD, while only preliminary results are available for other modalities. The evidence involving premanifest HD is much sparser. Key implications for clinical practice and future research are outlined and discussed.

Plain Language Summary

People with Huntington's disease can face psychological problems that affect their daily lives. One common issue is difficulties understanding what emotions other people feel. Scientists have studied this topic since they found the Huntington's gene in 1993, yet the full picture remains unclear. This review looked at 59 studies on emotion recognition in Huntington's disease published between January 1993 and January 2025 across four major databases. The results show that in people with manifest HD recognising angry, fearful, disgusted, and sad faces is often difficult. On the other hand, spotting happy or neutral faces is usually fine. A few experiments that used voices instead of faces showed similar weaknesses for disgust, fear, and anger in manifest HD, although happiness and sadness were less impaired. Evidence for understanding emotions from body postures, short stories, films, smells, or tastes is still limited. Some but not all people with premanifest HD also struggle with emotion recognition, probably because of early damage in frontal areas of the brains. These findings are important for researchers and people with HD alike, as they can help families, friends, caregivers, and patients respond better to daily challenges.

Introduction

Huntington's disease (HD) is a progressive neurodegenerative disorder linked to CAG repeat expansion in a mutated gene (Huntingtin) on the short arm of chromosome 4 [1]. It causes progressive basal ganglia damage, particularly involving the corpus striatum (caudate nucleus and putamen) [2]. This in turns leads to the development of motor impairments – such as chorea (involuntary movements), dystonia, bradykinesia, dysarthria, dysphagia, and rigidity [3]. The transmission mechanism is autosomal-dominant, meaning that affected individuals' children have a 50% probability of inheriting the gene [1]. Since 1993, genetic testing has been available to ascertain positive gene status [4].

The prevalence of HD in the UK is around 12.3 people per 100,000 [5], while the global pooled prevalence is estimated to be 4.88 per 100,000 [6]. The development of motor difficulties, which tends to occur around the age of 40 [1], is normally considered the onset of the condition, after which people with HD (pwHD) are considered 'manifest' or 'symptomatic'. People who have tested positive but do not yet experience movement issues are considered 'premanifest' or 'presymptomatic', while symptom-free individuals with a family history of the condition but no genetic testing are often described as 'at-risk'.

HD progression is also associated with a wide range of cognitive impairments [7]. These represent one of the earliest detectable clinical signs, with reduced speed of information processing being one of the strongest predictors of disease onset at the premanifest stage and disease progression at the manifest stage, particularly due to early involvement of the striatum [8]. Other common cognitive impairments involve reduced executive functions (e.g., planning, organisation, attention-shifting, self-monitoring, mental flexibility, and goal-directed behaviour; linked to disruptions to frontostriatal circuits), attention and automation (particularly evident on dual tasks, such as walking and talking at the same time), memory (mostly due to executive deficits rather than hippocampal disruption) and social cognition [7,9–14].

Among the problems involving social cognition, deficits of emotion recognition – defined as the ability to perceive and interpret affective information correctly in and from others [15,16] – have been consistently described in pwHD, especially regarding facial recognition of negative emotions such as anger, fear, and disgust [17–19]. While a number of recent reviews have addressed broader social cognition issues in this population [20–22], only one systematic review has so far focused on emotion recognition deficits in pwHD specifically [23]. This included studies between 1993 and 2010 and concluded that the vast majority of studies showing emotion recognition impairments in HD used visual tasks involving the identification of emotions from facial stimuli, with the evidence based on different types of stimuli (e.g., emotional body language) [24,25] or other sensory modalities (e.g., voices, tastes, and odours) [26,27] being much scarcer. Similarly, the evidence around impairments in people with premanifest HD appeared to be sparser, with fewer studies involving this population and more inconclusive findings [23].

Consequently, as 15 years have passed since the previous major review, and the literature and assessment of emotion recognition has progressed over this time period, an update is now warranted. In addition, the review by Henley and colleagues [23] only included comparisons between manifest or premanifest individuals and a control group of healthy volunteers, excluding any studies comparing single pwHD groups with published normative data or between-group comparisons across different HD stages. Thus, the present study aimed to fill these gaps by providing a more comprehensive systematic review of emotion recognition in Huntington's disease since the discovery of the Huntingtin gene.

Methods

For the purpose of this study, a systematic review approach was adopted, following the latest Preferred Reporting Items for Systematic reviews and Meta-Analyses (PRISMA) guidelines [28]. More specifically, the present review was guided by the following research questions:

1. Is emotion recognition impaired in people with HD?
2. Are impairments different between premanifest and manifest stages?

3. Are impairments different across emotions?
4. Are impairments different across stimuli and/or sensory modalities?
5. What methods are used to assess emotion recognition in people with Huntington's disease?

Due to the high level of heterogeneity in study designs, operationalisation of variables, methods of measuring emotion recognition, and reporting of key aspects of findings (e.g., effect sizes) highlighted within the literature by previous comparable reviews [23,29], a meta-analytic approach was not considered viable for this study.

Search Strategy

A comprehensive search was performed from January 1st 1993 (year of the discovery of the Huntingtin gene) to January 1st 2025, using a combination of free text terms across four major databases: Academic Search Ultimate (ASU), CINAHL, MEDLINE, and PsycINFO. Hand searches were also performed in reference lists of included studies in order to identify further relevant citations. Guidance was provided by a subject specific librarian regarding the search terms and strategy. The search logic grid as well as the full search strategy can be consulted in the Supplementary Information.

Inclusion Criteria

To be included in the present review, studies had to a) include adults (aged 18 and over) with manifest or premanifest HD; b) report quantitative data allowing for the analysis of group differences or effects sizes; c) compare HD gene carriers (either manifest or premanifest) to another group and/or normative data on tasks requiring the identification of human emotions based on stimuli from any sensory modality (e.g., visual, auditory). The decision to include only studies involving adult participants was made in light of the evidence that the biological and clinical manifestations of juvenile-onset HD differ significantly from those of adult-onset HD [30].

Quantitative observational or experimental studies with a cross-sectional, between-subjects, or quasi-experimental design were included in the present review. Due to

financial limitations, only studies published fully in English were considered eligible. No geographical limits were applied to the database searches for published literature.

Studies not adhering to the concept under investigation, not published in full in the English language, involving participants aged 17 or below, or involving and/or relating to animals were excluded. Systematic reviews, reviews, commentaries, editorials, letters, and qualitative studies were also excluded. Grey literature was not included to ensure all evidence had undergone a formal peer review process and adhered to the highest level of scientific scrutiny.

Studies primarily focused on stimulus congruence, reaction times, or eliciting emotion expressions or experiences were not included; when studies mixed these with more traditional emotion recognition tasks, the recognition results were still included when available, even if they were not the main focus of the investigation.

Study Selection

Results from searches of electronic databases were imported into a reference management software, where duplicate citations were removed. Studies and guidelines were selected using the eligibility criteria described above. In the first phase, all titles were screened by two reviewers, and those that clearly did not meet the inclusion criteria were excluded. In the second stage, all abstracts and full text articles were screened for eligibility by two reviewers; in case of disagreement, a third reviewer provided a further level of scrutiny.

Data Extraction

Selected data items were coded and extracted into an Excel sheet. All data were extracted by three reviewers and double checked by a further three to ensure accuracy. Data items for extraction included authors, year of publication, country of origin/study, type of study (e.g., design), key characteristics of the population (e.g., number, gender, sample power, context of recruitment, confirmation of HD status, premanifest or manifest, disease stage), types of emotion recognition investigated, emotion recognition measures adopted, and relevant results (e.g., means, SDs) and conclusions.

Data Synthesis

The extracted data were initially synthesised by two reviewers, who produced a preliminary narrative summary. This was then checked for consistency by a further two reviewers, before being revised by the whole research team during the manuscript write-up.

Quality Assessment

The appraisal of quality and risk of bias of the studies included in the review was first performed by two reviewers independently, who then cross-checked the results. In light of the design of the majority of the studies (i.e., non-interventional, between-groups) as well as the heterogeneity of adopted methods, the process criteria did not include standardised tools but were instead informed by previous comparable reviews [23,29]. [23,29] More specifically, these focused on a technical appraisal of elements such as sample size and power analysis, demographic data, control groups, the nature of stimuli and their presentation, response options, and potential confounding variables. Any disagreements between the two reviewers were solved by collective discussions with the whole Research Team.

Results

From an initial return of 9735 citations across MEDLINE, PsycINFO, CINHAL, and ASU, 6577 were retained following screening for duplicates and articles which were peer reviewed and fully published in English. From these, 6478 citations were excluded after screening by title and abstract, leaving 99 full-text articles to be considered for inclusion. Following full-text screening, 59 studies were eventually included in the present review. Figure 1 illustrates the PRISMA diagram for the selection of the studies, while their main characteristics are summarised below and outlined in Tables 1-8. See Supplementary Information for the list of citations excluded following full-text screening.

Design

The vast majority of the investigations (55, 93.2%) adopted a cross-sectional between-groups design comparing people with HD at different stages and some form of control group. Three papers adopted a single case design [31–33], while only one study carried out a case series of five individuals [34].

Countries

Most of the included studies were carried within high income countries, with Western Europe alone contributing to just over 70% of the evidence base. With regards to specific countries, the United Kingdom was the most represented (16, 27%), followed by France (7, 11.9%), Australia (6, 10%), and Germany and Denmark (4 studies/6.8% each). North America only accounted for four studies – three from the United States (5%) and one from Mexico (1.7%) – while China, Israel, and Argentina were only represented by a study each [35–37].

Sample Size

Sample sizes for pwHD ranged from single case studies to large multicentre investigations enrolling over 400 participants (e.g., the TRACK-HD cohorts) [38–40]. Control samples were similarly variable, ranging between one and 217 participants, the latter from a large international online study [11]. However, median group sizes were modest (i.e., around 18 for pwHD and 20 for controls), indicating predominantly small-to-medium scale studies consistent with the rarer nature of HD.

Type of HD Participants

Just over half of the included studies (39, 59%) recruited only pwHD at the manifest stage, while around 11 (19%) only recruited premanifest individuals. Another fifth (13, 22%) enrolled both manifest and premanifest pwHD.

Demographic Information

The mean age of pwHD was around 47 years old; of these, manifest groups, as expected, tended towards the late 40s or early 50s, while premanifest groups were typically five to seven years younger. The whole range of means spanned from age 30 in premanifest

samples to around 61 in manifest ones. Control groups showed comparable distributions, with means between 31.7 years [41] and 65.5 years [42].

HD samples were on average predominantly male (i.e., 44% female), while control groups were slightly more balanced (51% female). Where reported, HD groups showed a median of around 12.7 years of formal education, with a range between 4.1 years [39] and 20 years [32]. Controls showed almost identical figures (median: ≈ 12.5), though the upper end of the range was lower (16 years) [43]. Mean IQ levels for HD participants fell within the average range (108), ranging from 99 [36] to 117 [33]. The mean of controls was similar (107), with a range between 104.5 [44] and 109 [45]. Neither type of sample ever exceeded the superior range on average.

Sensory Modalities

Out of the 59 included studies, 56 (94.9%) included tasks investigating the visual modality (e.g., faces, eyes, visual vignettes). Auditory stimuli were a distant second, with only nine studies adopting them. Olfactory and gustatory stimuli were adopted concurrently by only two studies (3.4%) [26,27], while multimodality stimuli (i.e., mixing one or more sensory modalities, such as audiovideo) were adopted in four investigations [31,35,46,47]. No study investigated emotion recognition through tactile stimuli. Table 1 summarises the results by study, while Figure 2 illustrates the distribution of sensory modalities across all included studies.

Of the 56 studies adopting tasks investigating visual emotion recognition, an overwhelming majority (48, 85.7%) used facial stimuli, while eyes alone accounted for 24 studies (42.9%). Only around 17% of all included studies investigated other visual emotion recognition modalities, such as body language [24,25,48], visual vignettes [27,49,50], written vignettes [50,51], words [27], and audioless videos [32,52]. Figure 3 outlines the distribution of specific types of stimuli across the studies which explored the visual modality.

Technical Appraisal

As mentioned above, the process criteria for the technical appraisal of the included studies were informed by previous comparable reviews [23,29]. The details of the appraisal are outlined below, while Table 2 provides a summary for each study.

Control Samples

Almost all included studies (56, 94.95%) compared pwHD with some form of control group, with 44 comparing people with manifest HD against controls and 20 premanifest individuals against controls. Only single studies compared manifest [34] or premanifest [31] pwHD against standardised norms. Most included studies included healthy controls matched for age, gender and/or education. A smaller number of studies included gene-negative individuals as controls [35,53–55].

Longitudinal Designs

Only five studies adopted longitudinal designs [31,42,53,55,56], following participants for up to six years. Of these, one was a single case study [31], and the remaining ones adopted a between-subjects design. Two enrolled either only premanifest [31,55] or manifest [42,56] individuals, while only one study [53] recruited both.

Power Analysis and Sample Size Considerations

The majority of the studies relied on convenience samples, with only four reporting an a priori power calculation [11,37,57,58]. Three of these also contributed to the 11 investigations (18.6 %) that included some type of explicit consideration or justification around the adopted sample size [11,37,58].

Data Normality

Fewer than half of the included studies (24, 40.7%) reported considerations around data normality, suggesting that violations of test assumptions may have gone unrecognised in other investigations.

Family-Wise Error Rates (FWER)

Just over half of the investigations (34, 57%) clearly addressed family-wise error rates (FWER) when running multiple comparisons, either by applying corrections such as Bonferroni or False Discovery Rate (FDR), or by providing a rationale for not applying any adjustment (e.g., excessive conservativeness with smaller sample). Thus, the risk of false-positive findings may have been increased in the remaining 25 studies which did not report any consideration for FWER.

Clinical Characteristics

The reporting of some of the essential clinical characteristics of pwHD was quite inconsistent across studies. While CAG-repeat lengths were reported by a large majority (41, 69.5%), only around a quarter of the included studies (16, 27.1 %) reported actual or estimated IQ scores.

Effect Sizes

Less than one third of the included studies (18, 30.5%) reported standardised effect sizes when presenting their results, meaning that the current literature severely limits insight into clinical significance. However, the fact that the studies that did report effect sizes were published more recently appears to suggest a potential trend improvement in this regard.

Emotion Recognition Measures

No measures of emotion recognition specifically developed for or validated with pwHD are currently available. Thus, all included studies relied on measures developed for other populations or custom tools. While efforts have been made to describe the most common measures based on other populations as clearly as possible, custom tools as well as some of the lesser-known tests often feature detailed descriptions which require their specific context of use to allow for a full characterisation. Readers are therefore invited to consult the relevant citation to learn more about these.

Visual Measures

The investigation of visual emotion recognition relied heavily on face-based measures, with 26 studies adopting Paul Ekman's classic Pictures of Facial Affect (POFA) [59] – a stimulus set consisting of 110 frontal photographs depicting six basic emotions (anger, fear, surprise, happiness, sadness, and disgust), along with a neutral expression. It should be noted that studies adopting the Ekman 60 Faces Test (Ek-60F) [59] were also counted as using the POFA since the test draws from the same 110-picture stimulus set.

The second most common measure, adopted by 23 studies, was the Reading the Mind in the Eyes Test (RMET) [60], which focuses on the recognition of emotional expressions from photographs of eyes alone. The Emotion Hexagon Test (EHT [61]; characterised by facial images morphing within a hexagonal shape with expressions blending two different emotions) [61], was adopted by 14 investigations, while non-standardised custom tasks were developed ad hoc by nine studies. The Karolinska Directed Emotional Faces set (KDEF) [62], a set of 4900 pictures delivered by 70 individuals displaying 7 different emotional expressions from 5 different angles, was used by seven studies – on two occasions in a custom combination with the POFA [63,64]. The Facial Expressions of Emotions: Stimuli and Tests (FEEST; a computerised, updated version of the POFA) [65] was adopted by six investigations, while visual components of the Florida Affect Battery (FAB)[66] – a test measuring facial emotional discrimination, emotional prosody discrimination, and facial-auditory cross-modality tasks – was adopted by one study.

Some of the less commonly adopted measures (i.e., by fewer than five studies) included the Amsterdam Dynamic Facial Expression Set (ADEFS) [67], the Affective Picture System (IAPS) [68], the Manchester Face Set (MFS) [69], the Bochum Emotional Stimulus Set (BESST) [70], the Jerusalem Facial Expressions of Emotion (JeFEE) [71], the Emotion Recognition Task (ERT) [72], and the Advanced Clinical Solutions – Affect Naming (ACS-AN) [73].

Auditory Measures

Studies exploring auditory emotion recognition relied predominantly on custom tasks based on non-verbal sounds, with only one standardised measure, the emotional prosody discrimination component of the FAB [66], being adopted by one investigation [33].

Olfactory and Gustatory Measures

No standardised measures were adopted by the two studies investigating olfactory and gustatory emotion recognition, with both using custom odour and taste emotional labelling protocols [26,27].

Multimodality Measures

Almost all the six studies which explored emotion recognition across multiple modalities used tasks based on video vignettes, with the most common being The Awareness of Social Inference Test – Emotion Evaluation Task (TASIT-EET) [74] and the Amsterdam Dynamic Facial Expression Set videos (ADEFS) [67]. One investigation [46] also adopted a custom video vignettes measure, while another [31] used a composite score based on a combination of faces and non-verbal sounds from the FAB [66].

Emotion Recognition Findings

The main findings on all emotion recognition modalities are outlined below, divided by type of comparison. Due to their stimulus heterogeneity, the results for the visual modality are presented according to the specific type of tasks. For these, results from studies adopting larger participant cohorts (e.g., TRACK-HD) are also highlighted where relevant. Tables 3-7 summarise the findings for each modality, while Table 8 provides an outline of the main characteristics and key findings for of all included studies.

It should be noted that not all investigations within each modality explored all basic emotions (i.e., anger, fear, disgust, happiness, neutral, sadness, surprise) and some studies carried out multiple comparisons per modality using different measures. As a consequence, the number of studies reported for each emotion below may not match with the total number of studies in each modality.

Facial Emotion Recognition

Manifest v. Controls. When comparing manifest individuals against matched controls on facial emotion recognition tasks, impairments in pwHD were reported by over 60% of the studies adopting a total or composite emotion recognition score (7/11). On tests of specific emotions, significantly poorer facial emotion recognition for negative

emotions (e.g., anger, sadness) was also reported consistently in the HD group. More specifically, over three-quarters of the studies that measured anger (29/35), fear (26/35) and disgust (30/35), and more than 60% of the ones exploring sadness (22/34), reported significant poorer performance in the HD group – including all the studies from the TRACK-HD multicentre cohorts [38–40]. Recognition of surprise was also found to be impaired in more than half of the investigations (19/32); however, no impairment was reported in one of the TRACK-HD studies [38]. Finally, performance on facial stimuli for happiness was largely intact across studies, with just over one in five reporting impairments (8/35); the same was observed for neutral stimuli, albeit based on much fewer investigations (4/10). However, it should be noted that significant deficits for both happiness and neutral items were found in two of the TRACK-HD studies [39,40].

Manifest v. Norms. Only one study compared manifest pwHD to published norms on facial emotion recognition [34], finding a significant impairment using the total Facial Affect Recognition Scale of the FAB [66].

Premanifest v. Controls. When comparing premanifest individuals against matched controls, impairments in facial emotion recognition were reported less frequently. None of the three studies adopting a facial composite score found significant differences between groups [52,53,75]. Similarly, less than half of the emotion-specific investigations reported deficits for anger (5/12), fear (3/10), and disgust (5/12). Only one study reported impairments for sadness [76], while the TRACK-HD premanifest cohort evaluated by Labuschagne et al. [39] was one of the only two investigations (along with Sprengelmeyer and colleagues [55]) to report deficits of recognition of surprise. Facial recognition of happiness and neutral faces was entirely preserved in people with premanifest HD across all studies.

Manifest v. Premanifest. Compared to premanifest individuals, people with manifest HD were found to be impaired in most studies adopting a facial composite or total score (2/3). The two studies which compared participants on specific emotions [45,54] both found significant impairments of recognition of anger, fear, and disgust in manifest pwHD; however, only Henley et al. found deficits for recognition surprise and happiness, while only Milders and colleagues found a deficit for sadness. The latter was also the only

study to compare manifest and premanifest people on neutral facial stimuli, finding no significant difference between groups [45].

Premanifest v. Norms. Only one single case study compared the facial emotion recognition performance of a premanifest individual against published norms [31], finding a significant discrepancy with normative data using the total Facial Affect Recognition Scale of the FAB [66].

Eyes Emotion Recognition

Manifest v. Controls. Eleven out of 12 investigations which compared manifest individuals to matched controls on tasks involving eye-based emotion recognition adopted the RMET [60], which only yields a total score. Of these, 10 found a significant impairment in pwHD. The only study which measured specific emotions using a custom eyes-based task [77] found significant impairments for recognition of fear, disgust, and sadness, but not of anger, happiness, or surprise.

Premanifest v. Controls. All the nine studies which compared premanifest individuals against controls on eyes emotion recognition adopted the RMET. Unlike with manifest participants, however, the results of these investigations were less consistent, with more than half of the studies (5/9) finding no significant impairment in premanifest pwHD – including an international online study enrolling 117 participants across Italy and the UK [11].

Manifest v. Premanifest. The three studies comparing premanifest and manifest individuals on eyes emotion recognition all adopted the RMET and found significantly poorer performance in the manifest group than the premanifest [53,75,78].

Body Language Emotion Recognition

Manifest v. Controls. Two studies used emotional body language pictures with hidden facial expressions to compare manifest individuals with matched controls. De Gelder et al. [25] developed a custom task and found impairments for anger and neutral stimuli, but not for fear and sadness. Zarotti et al. [48] instead adopted 70 frontal body

language pictures from the standardised Bochum Emotional Stimulus Set (BESST) [70] and found impairments for recognition of fear, sadness, and neutral stimuli, but not of anger, disgust, happiness, or surprise.

Premanifest v. Controls. Only one study explored emotional body language recognition in premanifest pwHD, comparing them to matched controls [24]. The results, based on a custom task, showed no significant group differences for anger, disgust, or sadness.

Other Visual Emotion Recognition

Manifest v. Controls. Three studies adopted visual vignette stimuli from the International Affective Picture System (IAPS) [68] – a standardised set of emotionally evocative pictures including both human (e.g., faces) and non-human (e.g., rooms) stimuli – to compare manifest pwHD with matched controls. On one hand, Eddy et al. [50] found impairments for fear, but not disgust and happiness. On the other, Hayes et al. [27] found that manifest individuals showed a deficit for disgust, but not fear, happiness, or neutral stimuli. These results were corroborated by a later study by the same group, which found again significant impairments for disgust but none for anger, fear, or sadness [49]. Written emotional stimuli, whereby participants had to read emotional vignettes or words (and hence rely on vision), were also adopted by three studies [27,50,51]. Among these, however, only Calder and colleagues [51] found a significant difference between groups, specifically for anger. Finally, Larsen and colleagues [52] adopted an audioless version of the videos from the TASIT-EET [74] and found a significant impairment in manifest pwHD on the measure's total score. Similarly, Caillaud et al. [32] reported a case study in which a manifest individual was found to have no impairment based on the audioless video stimuli from the ADFES [67].

Premanifest v. Controls. An audioless version of the TASIT-EET [74] was used by Larsen and colleagues [52] to compare premanifest individuals with matched controls, finding no significant differences between groups on its total score.

Auditory Emotion Recognition

Manifest v. Controls. Each of the seven studies which used auditory emotion recognition tasks to compare manifest individuals against matched controls found a significant impairment for recognition of disgust, with fear and anger being a close second (6/7 and 5/7 studies respectively). On the other hand, recognition of happiness and surprise were found to be impaired in less than half of the investigations (2/5), and only one reported deficits of recognition of sadness [18]. None of the studies measured neutral expressions or adopted composite or total scores.

Premanifest v. Control. Only one study compared premanifest individuals to controls using a custom auditory task, finding no significant differences on a composite score [55].

Premanifest v. Norms. One study compared premanifest pwHD to published norms [31], reporting a significant impairment in the total Vocal Affect Recognition Scale of the FAB [66].

Olfactory and Gustatory Emotion Recognition

Manifest v. Controls. Only two studies investigated olfactory and gustatory emotion recognition in pwHD by comparing manifest individuals with matched controls on custom measures for both modalities. Hayes and colleagues [27] found a significant difference on the composite score for olfactory emotion recognition based on odorants, but not for gustatory stimuli (i.e., liquids). On the other hand, Mitchel et al. [26] found a significant impairment in the HD group on both olfactory (i.e., odorants) and gustatory (i.e., foodstuff) tasks.

Multimodality Emotion Recognition

Manifest v. Controls. The three studies which compared manifest individuals with matched controls on multimodal emotion recognition tasks (all based on video vignettes) yielded contrasting results. Philpott et al. [47] found a significant group difference using the full (i.e., both audio and video) TASIT-EET [74], whereas Caillaud et al. [46] did not find any significant impairments when adopting a custom video task ('Pierre

and Marie'). Similarly, Baez et al. [35] found no group differences based on the TASIT-EET total score or any of its specific emotional items.

Premanifest v. Norms. Burke et al. [31] reported a case study in which a premanifest individual was found to be impaired on the multimodality component (i.e., matching emotional prosody with emotional faces and vice versa) of the FAB [66].

Emotion Recognition Associations

Demographic Associations

Johnson et al. [38] found reduced emotion recognition performance was associated with increasing age, lower estimated IQ, and lower educational level. There appeared to be no gender effect in participants in most studies, although Johnson et al. [38] found females performed better than males. Larsen et al. [52] and Rees et al. [18] found emotion recognition deficits even after controlling for social and environmental factors, suggesting these may not be contributing variables.

Clinical Associations

A number of studies investigated the potential correlation between performance in emotion recognition and clinical variables such as CAG length, CAG-Age-Product (CAP) scores, or estimated time to disease onset in premanifest individuals. Mason et al. [79] and Larsen et al. [52] reported higher CAG-Age-Product (CAP) score and estimated time to disease onset were associated with worse emotion recognition performance. However, Croft et al. [80] reported that CAG repeat length was not associated with emotion recognition scores. Similarly, Ille, Schäfer, et al. [81] and van Asselen et al. [82] found no correlations between emotion recognition and CAG repeats, symptom duration, and the Total Motor Score (TMS) of the Unified Huntington's Disease Rating Scale (UHDRS) [83].

However, other studies did find that the UHDRS TMS correlated with emotion recognition from faces [37] and body language [25], while Total Functional Capacity scores from the same scale were positively correlated with body language recognition [48]. Furthermore, emotion recognition was found to predict decline in functional capacity over a six-year

follow up, independently of executive function, depression, and baseline disease severity [75]. However, Eddy et al. [84], Tinkler et al. [63], and Ille, Holl, et al. [81] found no correlations between disease duration, age of motor onset, and disease burden. Finally, higher levels of insulin-like growth factor 1 (IGF-1) were found to be significantly associated with better facial emotion recognition performance in a small sample of manifest pwHD [58].

Cognitive Associations

Across the included studies, some found associations between cognitive status or executive function and emotion recognition in HD participants [38,42,45,47,77], while others did not [37,81,82,85]. In several investigations, single measures of executive functioning correlated with emotion recognition [49,78]. However, Baez et al. [35] found no correlation between emotion recognition and executive functioning, and a longitudinal six-year follow up study showed that emotion recognition and functional decline were separate from executive functions [75].

Some studies also considered whether a deficit in basic facial recognition was associated with performance on emotion recognition tasks. In this regard, the evidence appears again inconsistent, with some findings showing that better face recognition predicted more accurate emotion recognition [18,38,45], but others reporting no association between facial recognition and emotion recognition [49,51]. Furthermore, after controlling for impairments on face matching in their analyses, Calder et al. [51] and Rees et al. [18] found the effect of poor recognition of negative emotions remained significant for HD participants.

Psychological Associations

Some studies controlled for anxiety and depression levels when analysing emotion recognition performance. Hendel et al. [75] found deficits even after controlling for depression in regression analyses. However, Croft et al. [51] and Johnson et al. [38] found no association between emotion recognition performance and levels of depression. In addition, difficulties in emotion regulation showed no relationship with emotion recognition performance across two studies [11,48]. Results for apathy appear more contrasting, with Hendel et al. [53] reporting that apathy correlated with emotion

recognition, but no association was found in another study [64]. Finally, Baez et al. [35] found no correlation between emotion recognition and empathy in pwHD.

Medication Associations

Osborne-Crowley et al. [40] found no effect of taking antidepressants or antipsychotics on emotion recognition. However, in early-stage HD, Labuschagne et al. [39] found that neuroleptics were associated with worse recognition performance, while selective serotonin reuptake inhibitors (SSRIs) were associated with better scores on recognition tasks. Both investigations enrolled large samples from the TRACK-HD cohorts. Similar findings for antidopaminergic medications were also reported by a later study [58].

Cross-Modality Associations

Only two studies investigated potential associations of emotion recognition performances across multiple modalities in pwHD, both between the visual and auditory modality. Sprengelmeyer et al. [55] found significant positive correlations between the recognition of faces and voices expressing happiness, surprise, fear, and anger. However, no significant association was found for sadness and disgust. On the other hand, Hayes et al. [27] found a significant positive correlation between the recognition of vocal expressions of disgust and disgusting vignettes from the IAPS [68].

Neuroimaging Findings

Neuroimaging investigations were carried out by around one-third of the included studies (18/59). Most adopted structural methods – such as computerised tomography (CT) scans, volumetric magnetic resonance imaging (MRI), or voxel-based morphometry (VBM) – and hypothesised frontostriatal atrophy as the anatomical substrate of emotion recognition impairments in pwHD. More specifically, early CT results by Sprengelmeyer et al. [56] linked impaired recognition of negative facial expressions to caudate and frontal volume loss. Subsequent studies corroborated these findings by showing that poorer

emotion recognition performance was associated with reduced grey matter in the caudate, putamen, insula, and orbitofrontal cortex in manifest HD [54,86,87]. Similarly, two VBM analyses showed that subtle frontostriatal degeneration was significantly correlated with poorer affective social cognition in premanifest individuals [32,46]. In their VBM study, Gil-Polo and colleagues [58] found that reduced frontotemporal grey matter volume and cortical thinning were significantly associated with lower IGF-1 levels in manifest HD, which were in turn significantly correlated with lower emotion recognition performance. In addition, a diffusion-tensor imaging (DTI) study found that white matter disruptions in the corpus callosum, the frontal gyrus, right anterior cingulate cortex, insula and amygdala regions, cerebellum, and brainstem were significantly associated with poorer emotion recognition from faces and eyes in manifest pwHD [44].

Albeit less frequent in number, functional neuroimaging studies showed hypoactivation within areas such as the precuneus, anterior insula, anterior and posterior cingulate, and supramarginal gyrus in manifest HD during tasks involving emotion recognition [78,88]. Moreover, lower amygdala–fusiform connectivity and reduced activation of the superior temporal sulcus were observed in premanifest individuals when judging facial emotional expressions [76,79]. Finally, Novak et al. [89] found that reduced neural activity in premanifest pwHD – comparable to the other studies – could be partially distinguished based on the processing of three specific emotions (i.e., disgust, anger, and happiness).

Discussion

The present work aimed to provide a comprehensive systematic review of empirical studies on emotion recognition in people with HD over the three decades since the consistent identification of the HD gene through direct testing. From 9735 initial records, 59 studies were considered eligible for inclusion. Even when accounting for the more encompassing inclusion criteria of this work, the number of included studies has increased more than threefold compared to a previous review from 15 years ago [23], highlighting a huge surge in interest on the topic of emotion recognition within the HD

literature. Most included studies adopted a cross-sectional design, with only five following participants longitudinally and four reporting single cases or case series. Around 70% of the evidence came from Western Europe, and especially the United Kingdom, while other continents were much less represented. Sample sizes ranged from one to over 400, with median values around 20.

The results in people with manifest HD showed robust evidence of deficits of recognition of negative emotions from facial cues, with over 75% of relevant studies consistently reporting impairments for recognition of anger, fear, and disgust, and more than half for sadness and surprise. Facial recognition of happiness and neutral expressions appears instead to be more preserved, although some notable exceptions were two of the studies based on the large TRACK-HD cohorts [39,40]. On tests focused on eye stimuli, 90% of studies with manifest individuals reported evidence of global emotion recognition impairments, while one also reported specific deficits for fear, disgust, and sadness [77]. Auditory studies, although sparser than facial ones, all show consistent deficits for the recognition of disgust, fear, and anger, while happiness and sadness appeared less affected. Only preliminary evidence is currently available for deficits of emotion recognition from body language [25,48], visual and written vignettes [27,50,51], audioless videos [52], multimodal video vignettes [47], olfactory tasks based on odorants [26,27], and gustatory tasks based on foodstuff [26].

On the other hand, the evidence involving people with premanifest HD is currently much less consistent, with no significant differences found with matched controls on any composite or total scores of facial emotion recognition. When testing for specific emotions, impairments in recognition of anger, fear, and disgust were reported in less than 40% of relevant studies, while only isolated investigations reported issues with sadness [76] and surprise [55]. Facial recognition of happiness and neutral expressions was preserved across all premanifest studies. Findings involving tasks based on eye expressions were mixed, with around half showing no overall impairments, including a large international online sample [11]. Studies exploring recognition across other sensory modalities in premanifest HD were severely lacking, with isolated contrasting findings

available on auditory tasks [33,55], only single norm-based evidence for multimodal deficits [31], and no investigation available for olfactory or gustatory tasks.

Across both manifest and premanifest participants, older age, lower education or IQ, and greater motor impairments (as measured by the UHDRS-TMS) were linked to poorer emotion recognition, while gender or CAG repeat length showed weak or no associations. Associations with medications appear tentative, with some evidence suggesting that neuroleptics may lower recognition performance, while SSRIs may improve it. Only two investigations looked into associations of emotion recognition deficits across modalities, particularly visual and auditory, finding contrasting results [27,55]. Cognitive abilities such as executive functioning and basic facial recognition skills were associated with emotion recognition only in some studies, showing an overall inconsistent pattern which warrants further investigation due to the wide range of cognitive issues pwHD may experience at all disease stages [7,90]. Similarly, the exploration of associations between psychological difficulties and emotion recognition performance showed mixed results, with only sparse evidence available for anxiety, depression, and apathy. This represents again an issue which needs further attention, especially in light of the wide range of (often unrecognised) psychological difficulties which are experienced by people affected by HD [91–93] and the current severe lack of psychological support available for this population [94,95]. Finally, structural and functional neuroimaging studies consistently associated poorer emotion recognition with frontostriatal atrophy, white-matter disruption, and hypoactivation of large cortico-subcortical brain networks in both manifest and, more subtly, premanifest participants. These findings also appear consistent with neurobiological evidence that loss or disruption of hypothalamic neuropeptides such as oxytocin and vasopressin in pwHD may be implicated with anormal frontostriatal activation as well as reduced emotional processing and recognition [96–98].

Overall, the findings from this review highlight that impairments of emotion recognition in pwHD likely follow a progressive pattern which may not always be equally apparent across disease stages. Before motor onset, most premanifest individuals are still able to recognise emotions effectively from faces and eyes, but a small subset may show early subtle difficulties with specific emotional cues, although with patterns that are yet to be

clearly evidenced. These issues appear to be linked with the early involvement of frontostriatal areas which are essential for social cognitive tasks and are consistent with the other early cognitive difficulties which can often become evident before motor phenoconversion [8,21]. At the manifest stage, deficits become significantly more apparent across visual (faces, eyes, body language) and auditory modalities, and may also present in olfactory, gustatory, or multimodal tasks. In contrast to some of the earlier HD literature, which suggested emotion recognition impairments in affected individuals were selective for disgust [99], it is now clearer that these difficulties predominantly affect all negative emotions, while relatively sparing the recognition of positive affect (e.g., happiness) and neutral stimuli. When compared with premanifest individuals, manifest pwHD are also likely to show significantly poorer emotion recognition performance across the board.

Implications for Clinical Practice

A number of important clinical implications can be drawn from this review. Since the evidence indicates that, similarly to other cognitive skills, emotion recognition abilities in HD may deteriorate early in the premanifest stage and become more pronounced as the disease evolves, routine assessments of facial emotion recognition (as well as other modalities as further evidence accrues) may support the disease staging process and act as a potential cognitive biomarker of disease progression. This idea was already suggested by Henley and colleagues in their previous review 15 years ago [23] and now – with a three times stronger body of evidence – it should receive renewed attention in HD clinical practice. With regards to facial recognition measures, Paul Ekman's classic Pictures of Facial Affect (POFA) [59] may be considered as first choice, particularly due to its reliability, rich evidence-base with pwHD and other conditions, and its availability in a variety of different delivery methods – e.g., pencil and paper or computerised in the form of the FEEST [65].

In addition, when performing medication reviews, HD clinicians should consider the potential effects of different pharmacological treatment options, such as those based on neuroleptics or SSRIs, on emotion recognition. This appears especially relevant

considering the potential negative impact of emotion recognition deficits on the social interactions and quality of life in HD [23].

Finally, evidence has shown that psychotherapy can play a pivotal role in improving emotional awareness as well as general social cognitive skills [100–102]. Therefore, as further evidence accrues on the potential association between other neuropsychological difficulties (e.g., anxiety, depression, executive dysfunction) and emotion recognition, the development of targeted cognitive and behavioural interventions for pwHD should be prioritised with the aim to improve social cognition and quality of life of affected individuals and their families [94,103].

Methodological Limitations and Future Directions

Although interest in the topic of emotion recognition in pwHD has clearly increased in the past few decades, several methodological limitations exist within the literature, which prevented the adoption of meta-analytic approaches and provide caveats on the conclusions of this review. The issues highlighted previously [23] around power, heterogeneity of methods, small sample sizes, and poor reporting of key covariates (e.g., CAG repeats, IQ) still persist to an extent – albeit with some signs of improvement. For example, the TRACK-HD study allows for the investigation of emotion recognition in large international cohorts of pwHD.

In future studies, significantly more focus is therefore needed on the inclusion of crucial methodological elements such as increased power, adjustment for multiple comparisons, consideration of data normality, and reporting of effect sizes. As the present article only included literature published in English, future reviews should also aim to include studies published in other languages.

Methodological heterogeneity may also help explain some of the conflicting findings observed across studies, particularly when investigating multiple emotions, as the way specific elements of affect or emotion are operationalised may vary significantly across measures. For instance, facial emotion recognition from front-facing images (e.g., the POFA) may be easier than when adopting angled pictures (e.g., the KDEF [62] or EHT [61]). However, such differences are rarely accounted for when total scores are reported.

Future studies should therefore aim to provide a more comprehensive characterisation of the subtler differences across measures which may have an impact on participants' results.

In addition, further investigations are strongly warranted on emotion recognition based on visual stimuli other than faces and eyes (e.g., body language, visual vignettes) as well as currently neglected sensory modalities – such as auditory, olfactory, gustatory, and multimodality tasks. This is even more relevant for individuals with premanifest HD, in order to address the current sparse nature of the literature involving individuals at this stage and should adopt some of the standardised measures which have shown preliminary significant results in this review (e.g., the verbal components of the FAB [66]) rather than rely on custom tasks.

Finally, the further development of standardised assessment batteries across all sensory modalities, and their specific validation with pwHD, remains a high priority to reduce the impact of methodological heterogeneity, improve ecological validity, and shed new light into the different facets of emotion recognition impairments in this population.

Conclusions

In manifest HD, facial recognition of negative emotions such as anger, fear, disgust, and sadness is consistently impaired, whereas happiness and neutral expressions are generally spared. A small number of auditory studies show consistent deficits for disgust, fear, and anger, while happiness and sadness appear less affected. Only preliminary evidence is currently available for deficits involving body language, visual and written vignettes, videos, and olfactory and gustatory tasks. The evidence involving premanifest individuals is currently sparser; however, studies suggest that sporadic areas of significant emotion recognition weakness may develop in some people prior to the onset of motor symptoms, particularly due to early degeneration of frontostriatal pathways and disruption of white matter tracts. Clinicians should consider routine assessments of emotion recognition to aid staging and inform new targeted interventions. Future research should also focus on adopting more adequately powered, longitudinal designs

using standardised and validated emotion recognition tests, with the potential to establish a gold standard for the assessment of emotion recognition in this population.

Declarations

Competing Interests

The authors have no competing interests to declare.

Authors' Contributions

All authors contributed to the study conception and design. Material preparation, data collection, and analysis were performed by all authors. The first draft of the manuscript was written by NZ and AS and all authors contributed to further versions of the manuscript. All authors read and approved the final manuscript.

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Data Availability

Datasets from the present work can be accessed from the authors upon request.

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Tables

Table 1

Sensory Modalities Across Included Studies

Study	Visual	Auditory	Olfactory	Gustatory	Tactile	Multimodality
11	■					
18	■	■				
19		■				
24	■					
25	■					
26			■	■		
27	■	■	■	■		
31	■	■				■
32	■					
33	■					
34	■					
35	■					■
36	■					
37	■					
38	■					
39	■					
40	■					
41	■					
42	■					
43	■					
44	■					
45	■					
46	■					■
47						■
48	■					
49	■	■				
50	■					
51	■	■				
52	■					
53	■					

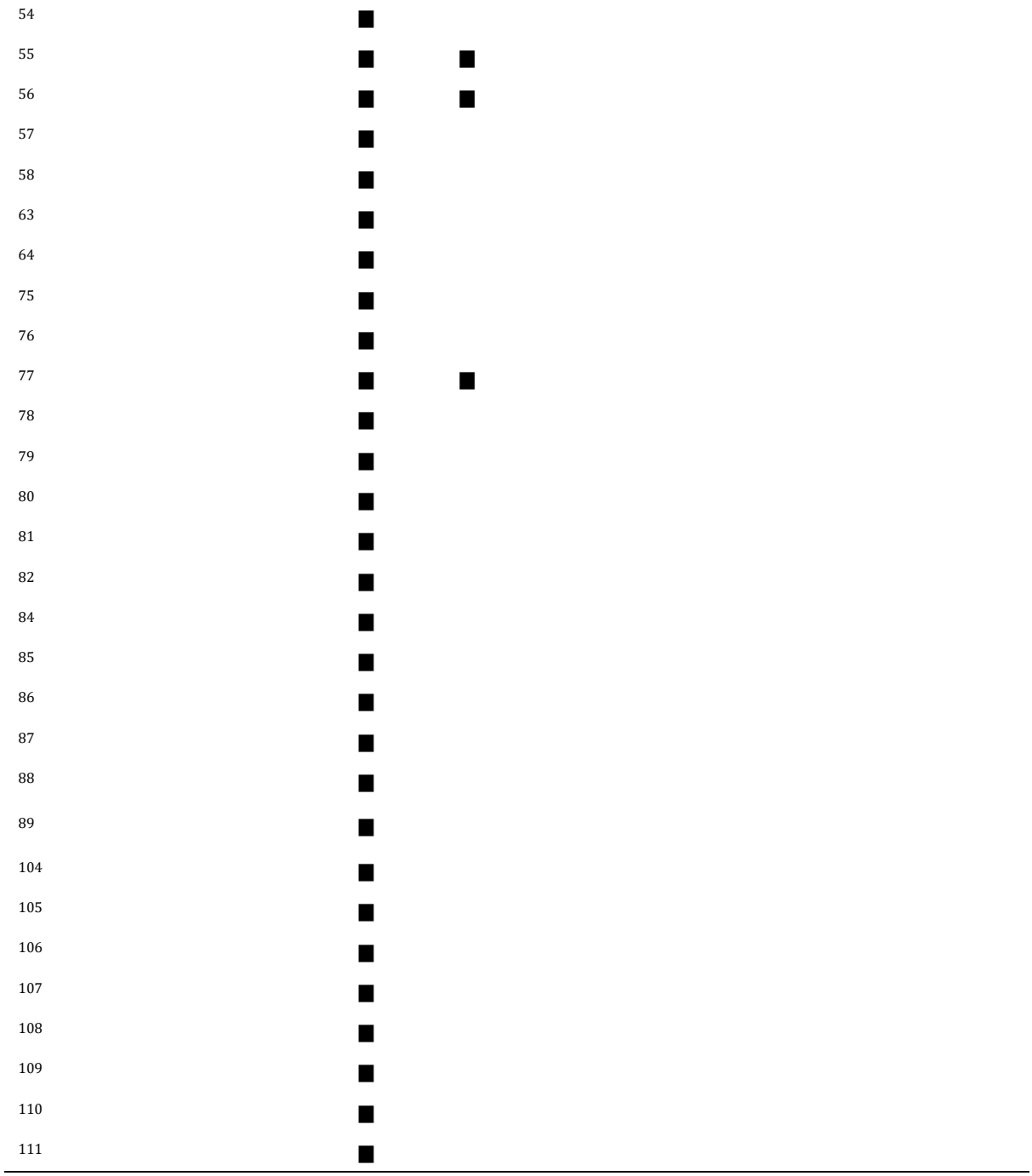
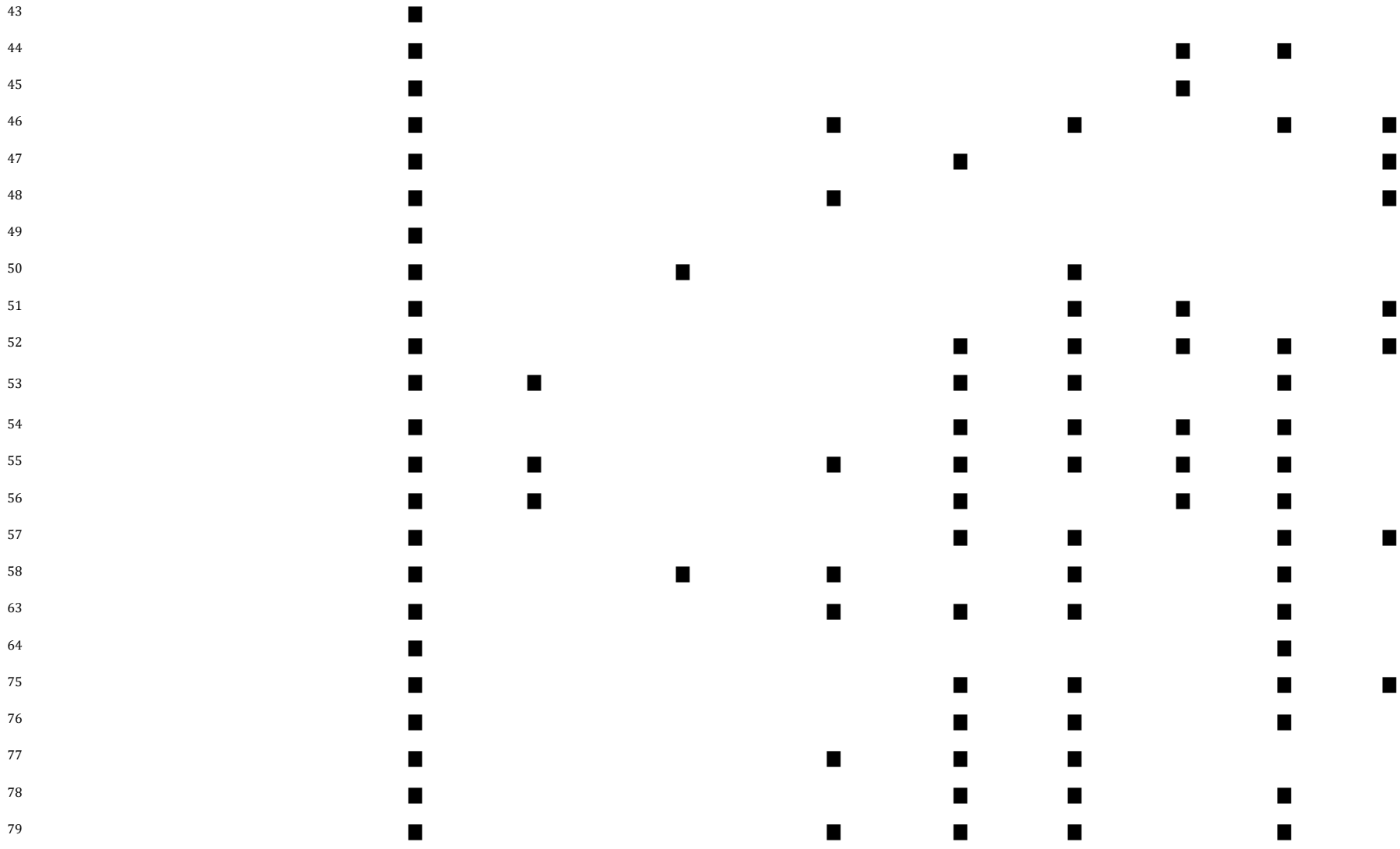


Table 2

Technical Appraisal of Included Studies

Study	Control present	Longitudinal design	Power analysis performed	Sample size considered	Normality tested	FWER addressed	IQ reported	CAG reported	ES reported
11	■		■	■	■	■			■
18	■				■			■	
19	■				■				
24	■							■	
25	■					■		■	
26	■								
27	■					■			
31		■					■	■	
32	■					■		■	
33	■						■	■	
34									
35	■					■	■		
36	■						■		
37	■		■	■				■	■
38	■			■	■	■	■	■	■
39	■				■			■	
40	■					■		■	■
41	■					■		■	■
42	■	■						■	■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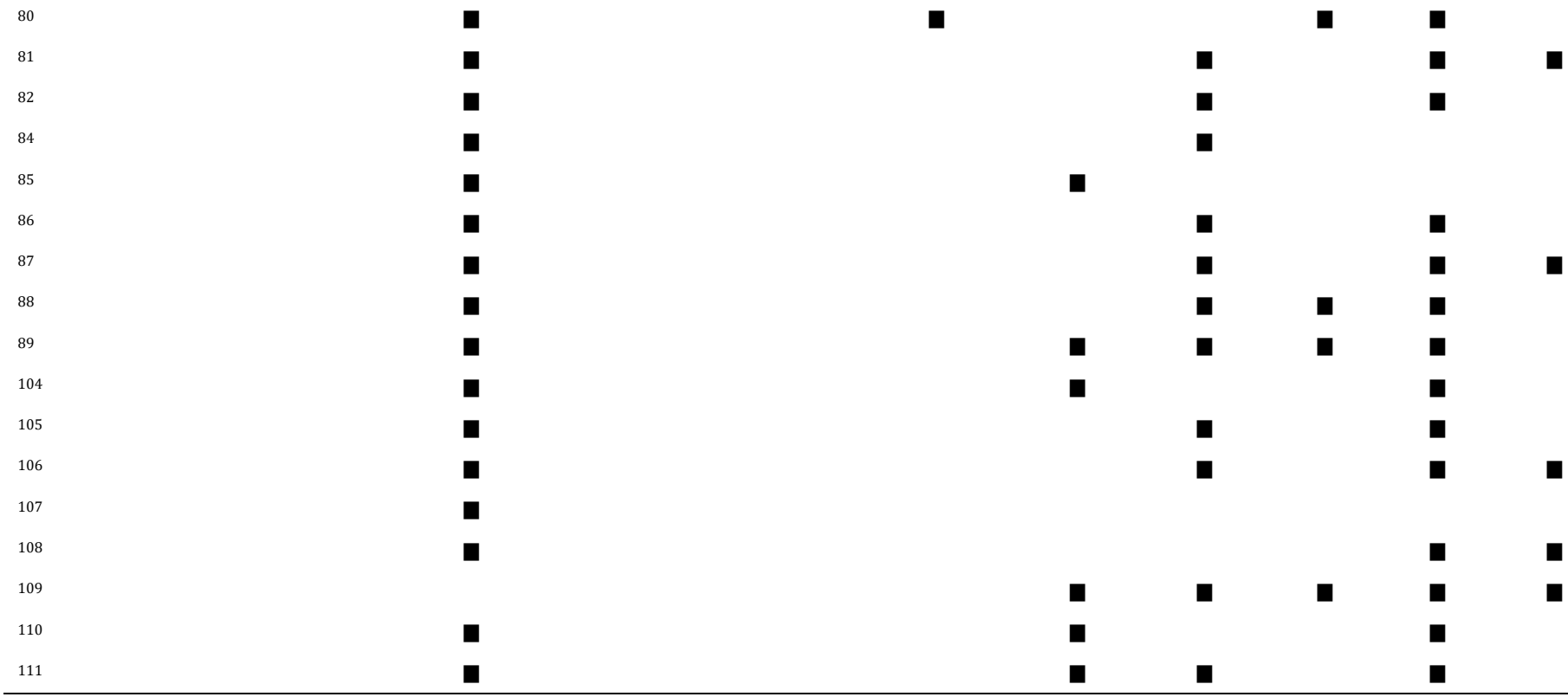


Note. ES = effect size; FWER = Family-Wise Error Rate.

Table 3

Findings for Visual Emotion Recognition

Comparison	Study	Stimuli	Measure	Anger	Fear	Disgust	Happiness	Neutral	Sadness	Surprise	T/C
Manifest/Ctrl	[104]	Eyes	RMET								◆
	[35]	Faces	POFA	○	◆	◆	○		○	○	
	[43]	Faces	POFA								○
		Eyes	RMET								◆
	[32]	Eyes	RMET								○
		Faces	ADEFS	○	○	○	○	○	○	○	○
		Audioless Videos	ADEFS	○	○	○	○	○	○	○	
	[46]	Faces	ADEFS								○
	[51]	Faces	POFA	◆	◆	◆	◆		◆	◆	
		Faces	EHT	◆	◆	◆	◆		◆	◆	
		Written Vignettes	Custom	◆	○	○	○		○	○	◆
	[80]	Faces	KDEF	○	○	○	○	○			
	[25]	Body language	Custom	◆	○			◆	○		
	[50]	Visual Vignettes	IAPS		◆	○	○				
		Written Vignettes	Custom		○	○	○				
	[84]	Eyes	RMET								◆
	[78]	Eyes	RMET								◆

[58]	Eyes	RMET								◆
	Faces	POFA								◆
[27]	Words	Custom		○	○	○	○	○		
	Visual Vignettes	IAPS		○	◆	○	○	○		
[49]	Faces	EHT	◆	◆	◆	○		◆	◆	
	Faces	FEEST	◆	◆	◆	○		◆		
	Visual Vignettes	IAPS	○	○	◆			○		
[75]	Faces	EHT								◆
	Eyes	RMET								◆
[53]	Faces	EHT								◆
	Eyes	RMET								◆
	Faces	POFA	◆	◆	◆	○		○	◆	
[81]	Faces	KDEF	◆	○	◆	○		○	◆	
[86]	Faces	KDEF	◆	○	◆	○		◆	◆	
[38]	Faces	POFA	◆	◆	◆	○	○	◆	○	
[106]	Faces	POFA	◆	○	◆	◆	○	◆	◆	
[39]	Faces	POFA	◆	◆	◆	◆	◆	◆	◆	
[52]	Faces	EHT								◆
	Eyes	RMET								◆
	Audioless Videos	TASIT-EET								◆

	[79]	Eyes	RMET								◆
	[45]	Faces	POFA	◆	◆	◆	○	○	◆	○	
	[107]	Faces	Custom	◆	○	◆	○		○	○	
	[40]	Faces	POFA	◆	◆	◆	◆	◆	◆	◆	◆
	[18]	Faces	MFS	◆	◆	◆	○		○	○	
		Faces	Custom	◆	◆	◆	○		○	○	
	[110]	Faces	POFA								◆
	[87]	Faces	KDEF	◆	○	○	○		○	○	
	[77]	Faces	POFA	◆	◆	◆	○		◆	◆	
		Faces	FEEST	◆	◆	○	○		○	○	
		Faces	MFS	○	○	◆	○		○	◆	
		Eyes	Custom	○	◆	◆	○		◆	○	
	[44]	Faces	POFA	◆	◆	◆	○		◆	◆	
		Eyes	RMET								◆
	[56]	Faces	POFA	◆	◆	◆	○		◆	◆	
		Faces	EHT	◆	◆	◆	○		◆	◆	
	[33]	Faces	EHT	○	◆	◆	○		○	○	
	[63]	Faces	POFA, KDEF	○	◆	◆	○		◆	○	
	[64]	Faces	POFA, KDEF	◆	◆	◆	◆		◆	◆	
	[42]	Faces	KDEF	◆	◆				◆		

	[82]	Faces	FEEST	◆	◆	◆	○	◆	◆	○		
		Faces	POFA	◆	◆	◆	◆	◆	◆	◆		
	[111]	Faces	EHT								◆	
	[36]	Faces	Custom	◆	◆	◆	○		◆	◆		
	[37]	Faces	JeFEE	◆	◆	◆	○		◆	◆		
		Faces	ADEFS	◆	◆	◆	◆		◆	◆		
	[48]	Faces	BESST	◆	○	◆	○	○	○	○		
		Body Language	BESST	○	◆	○	○	◆	◆	○		
Manifest/Norms	[34]	Faces	FAB								◆	
Premanifest/Ctrl	[24]	Faces	POFA	◆		◆	○		○			
		Body Language	Custom	○		○			○			
	[57]	Eyes	RMET								◆	
	[78]	Eyes	RMET								◆	
	[85]	Faces	POFA	○	○	◆	○	○	○	○		
	[75]	Faces	EHT								○	
		Eyes	RMET								○	
	[53]	Faces	EHT								○	
		Eyes	RMET								○	
	[54]	Faces	POFA	◆	○	◆	○		○	○		
	[88]	Faces	FEEST	○	○	◆	○		○	○		

	[105]	Faces	EHT	○	○	○	○		○	○		
	[39]	Faces	POFA	◆	◆	○	○	○	○	◆		
	[52]	Faces	EHT								○	
		Eyes	RMET								○	
		Audioless Videos	TASIT-EET								○	
	[79]	Eyes	RMET								○	
	[45]	Faces	POFA	○	○	○	○	○	○	○		
	[89]	Faces	POFA	○		○	○					
	[108]	Eyes	RMET								◆	
	[41]	Eyes	RMET								◆	
	[55]	Faces	FEEST	◆	◆	◆	○	○	○	◆		
	[82]	Faces	FEEST	○	○	○	○	○	○	○		
		Faces	POFA	○	○	○	○	○	○	○		
	[76]	Faces	ERT	◆	◆	○	○	○	◆	○		
	[11]	Eyes	RMET								○	
Premanifest/Manifest	[78]	Eyes	RMET								◆	
	[75]	Faces	EHT								◆	
		Eyes	RMET								◆	
	[53]	Faces	EHT								◆	
		Eyes	RMET								◆	

	54	Faces	POFA	◆	◆	◆	◆		○	◆		
	45	Faces	POFA	◆	◆	◆	○	○	◆	○		
	109	Faces	ACS-AN								○	
Premanifest/Norms	31	Faces	FAB								◆	

Note. ○ = no significant difference; ◆ = significant difference; ACS-AN = Advanced Clinical Solutions Affect Naming; ADEFS = Amsterdam Dynamic Facial Expression Set; BESST = Bochum Emotional Stimulus Set; EHT = Emotion Hexagon Test; ERT = Emotion Recognition Task; FEEST= Facial Expressions of Emotions: Stimuli and Tests (i.e., POFA + EHT); IAPS = Affective Picture System; JeFEE= Jerusalem Facial Expressions of Emotion; KDEF = Karolinska Directed Emotional Faces; MFS = Manchester Face Set; NimStim = NimStim Set of Facial Expressions; POFA= Pictures of Facial Affect; RMET = Reading the Mind in the Eyes Test; T/C = total or composite score.

Table 4

Findings for Auditory Emotion Recognition

Comparison	Study	Stimuli	Measure	Anger	Fear	Disgust	Happiness	Neutral	Sadness	Surprise	T/C
Manifest/Ctrl	[51]	Nonverbal Sounds	Custom	◆	◆	◆	○		○	○	
	[27]	Nonverbal Sounds	Custom	◆	○	◆			○		
	[49]	Nonverbal Sounds	Custom	○	○	◆			○		
	[18]	Nonverbal Sounds	Custom	○	◆	◆	○		○	○	
		Nonverbal Sounds	Custom	◆	◆	◆			◆	◆	
	[19]	Nonverbal Sounds	Custom	◆	◆	◆	◆		○		
	[77]	Nonverbal Sounds	Custom	◆	◆	◆	○		○	○	
	[56]	Nonverbal Sounds	Custom	○	◆	◆	◆		○	◆	
Premanifest/Ctrl	[55]	Nonverbal Sounds	Custom								○
Premanifest/Norms	[33]	Verbal Sounds	FAB								◆

Note. ○ = no significant difference; ◆ = significant difference; Ctrl = control.

Table 5

Findings for Olfactory Emotion Recognition

Comparison	Study	Stimuli	Measure	Anger	Fear	Disgust	Happiness	Neutral	Sadness	Surprise	T/C
Manifest/Ctrl	[27]	Odorants	Custom								◆
	[26]	Odorants	Custom								◆

Note. ◆ = significant difference; Ctrl = Control.

Table 6

Findings for Gustatory Emotion Recognition

Comparison	Study	Stimuli	Measure	Anger	Fear	Disgust	Happiness	Neutral	Sadness	Surprise	T/C
Manifest/Ctrl	[27]	Liquids	Custom								◌
	[26]	Foodstuff	Custom								◆

Note. ◌ = no significant difference; ◆ = significant difference; Ctrl = Control.

Table 7

Findings for Multimodality Emotion Recognition

Comparison	Study	Stimuli	Measure	Anger	Fear	Disgust	Happiness	Neutral	Sadness	Surprise	T/C
Manifest/Ctrl	[35]	Video Vignettes	TASIT-EET	◌	◌	◌	◌	◌	◌	◌	◌
	[46]	Video Vignettes	Custom								◌
	[47]	Video Vignettes	TASIT-EET								◆
Premanifest/Norms	[31]	Faces + Verbal Sounds	FAB								◆

Note. ◌ = no significant difference; ◆ = significant difference; Ctrl = Control; FAB = Florida Affect Battery; TASIT-EET = The Awareness of Social Inference Test - Emotion Evaluation Task.

Table 8

Main Characteristics of Included Studies

Study	Country	Design	HD Stage	HD Group N Age (M, SD) Gender (%F)	Ctrl Group N Age (M, SD) Gender (%F)	ER Modality	ER Stimuli	ER Measures	Key ER results
[11]	UK	Between-groups	Premanifest	117 37.38 (11.06) 70	217 40 (15.39) 51	Visual	Eyes	RMET	No significant difference observed between groups.
[18]	UK	Between-groups	Manifest	15 52.29 (9.41) 80	17 56.31 (8.92) 39	Visual Auditory	Faces Nonverbal Sounds	MFS Custom	HD group significantly worse than controls at recognising anger, disgust and fear, but not happiness, sadness, or surprise.
[19]	France	Between-groups	Manifest	14 51.29 (7.69) 42.8	15 46.80 (11.18) 46.6	Auditory	Nonverbal Sounds	Custom	HD group significantly worse than controls at recognising nonverbal sounds of happiness (achievement and pleasure), anger, disgust, fear, and pleasure.
[24]	Canada	Between-groups	Premanifest	21 48.3 (10.1) 57.1	27 49.2 55.6	Visual	Faces Body language	POFA Custom	HD group significantly worse at recognising anger and disgust in isolated facial expressions. No difference between HD group and controls in anger, sadness and disgust from body language.
[25]	France	Between-groups	Manifest	19 52.0 (9.1) 47.4	19 48.2 (8.0) 42.1	Visual	Body Language	Custom	HD group significantly worse than controls with full-body expressions. No impairment of fear or sadness.

[26]	UK	Between-groups	Manifest	8 NR NR	8 NR NR	Olfactory Gustatory	Odorants Foodstuff	Custom	HD group significantly worse at recognising disgusting odours compared to controls. HD group significantly less prone to recognise inappropriate food combinations as disgusting, but no differences in reaction to gustatory stimuli.
[27]	Australia	Between-groups	Manifest	14 54.6 (11.6) 40	14 51.3 (9.25) 40	Visual Auditory Olfactory Gustatory	Visual Vignettes Words Nonverbal Sounds Odorants Liquids	IAPS Custom	HD group significantly impaired at recognising disgust from written vignettes, but not fear happiness, sadness, or surprise. No impairment from words. Impaired recognition of anger and disgust from nonverbal sounds but not fear and sadness. General odour naming impairment. No significant impairment using taste.
[31]	Ireland	Single case study (longitud.)	Premanifest	1 50 0		Visual Auditory Multimodality	Faces Verbal Sounds Faces + Verbal Sounds	FAB	<u>Baseline</u>: Deficit on found on facial, verbal, and multimodal emotion recognition. <u>2-year follow-up</u>: No significant change.
[32]	France	Between-groups	Manifest	20 37.41 (8.96) 45	20 38.49 (10.14) 45	Visual	Faces Audioless Videos	ADEFS	No significant difference between HD and controls.
[33]	Germany	Between-groups	Premanifest	14 31 (1.8) 64.2	<u>Gene-negative:</u> 8 75 38.25 (14.51) <u>Matched:</u>	Visual Auditory	Faces Nonverbal Sounds		<u>Baseline and 6-month:</u> Faces: premanifest group significantly impaired on faces of disgust, but no other emotion compared to gene-negative; premanifest significantly impaired on faces of surprise, fear, disgust, and anger compared to matched controls. Sounds: no significant differences between

					37 36.23 (12.6) 54.1				premanifest and gene-negative groups on any emotions; no matched control comparison. <u>12-month:</u> Faces: premanifest group significantly impaired on faces of disgust and surprise, but no other emotion; no matched control comparison. Sounds: no significant differences between premanifest and gene-negative groups on any emotions; no matched control comparison. Recognition of faces and voices expressing happiness, surprise, fear, and anger correlated significantly in the premanifest group.
[34]	USA	Case series	Manifest	5 41 NR		Visual	Faces	FAB	Significant general impairments of emotion recognition in four out of 5 HD manifest HD patients.
[35]	Argentina	Between-groups	At-risk Manifest	<u>At-risk</u>: 19 29.2 (9.6) 68.4 <u>Manifest</u>: 18 43.8 (10.3) 50	<u>At-risk Ctrl</u>: 18 29.5 (10.2) 38 <u>Manifest Ctrl</u>: 18 43.2 (10.3) 66	Visual Multimodality	Faces	POFA TASIT-EET	At-risk and HD participants impaired in recognition of negative emotions with isolated faces (POFA) but normal in multimodality (TASIT-EET).
[36]	China	Between-groups	Manifest	6 44.8 (4.16) 33.33	16 45.19 (4.97) 37.5	Visual	Faces	Custom	HD group significantly worse than controls at recognising surprise, fear, sadness, disgust, and anger, but not happiness.
[37]	Israel	Between-groups	Manifest	21	21	Visual	Faces	JeFEE	HD group significantly worse than controls across all emotional stimuli. However,

				47.38 (13.20) 45	44.75 (13.98) 50			ADEFS	performance of in the HD group approached chance level when introducing more ecologically looking facial expressions.
[38]	Australia Canada USA	Between-groups	Premanifest Manifest	464 41.43 (9.63) 62.93	57 43.01 (10.13) 61.4	Visual	Faces	POFA	HD group significantly worse than controls on all negative emotions (anger, disgust, fear and sadness). No significant difference found for happiness, surprise, or neutral stimuli.
[39]	Canada France Netherlands UK	Between-groups	Premanifest Stage 1 Manifest Stage 2 Manifest	Pre-HD A: 61 41.0 (8.7) 52.5 Pre-HD B: 54 40.5 (9.2) 57.4 Stage 1 HD: 113 47.2 (10.3) 59.7 Stage 2 HD: 51.2 (8.7) 43.9	116 45.8 (10.3) 54	Visual	Faces	POFA	Premanifest HD groups significantly worse than controls at recognising anger, fear, and surprise. Nonsignificant trends for happiness, sadness, and disgust. Both Stage 1 and Stage 2 Manifest HD groups significantly worse controls across all stimuli: happiness, sadness, anger, fear, disgust, surprise, and neutral.
[40]	Canada France Netherlands UK	Between-groups	Manifest	Group 1: 43 48.43 (9.78) 47 Group 2: 67	107 46.13 (10.14) 54	Visual	Faces	POFA	HD group worse than controls on all emotional stimuli. HD participants with apathy worse than non-apathetic HD participants. Specific impairment in the recognition of happiness in HD participants with apathy compared to non-apathetic HD participants.

				48.20 (10.05) 61					
[41]	Italy	Between-groups	Premanifest Manifest	Premanifest: 20 34.9 (8.9) 50 Manifest: 40 45.3 (10.1) 50	Premanifest Ctrl: 40 31.7 (4.7) 50 Manifest Ctrl: Ctrl: 40 47.2 (7.2) 50	Visual	Eyes	RMET	Recognition significantly worse in premanifest and manifest HD compared to controls. Manifest group also significantly worse than premanifest.
[42]	Italy	Between-groups	Manifest	12 61.08 (11.90) 31	11 65.45 (10.34) 27.3	Visual	Faces	KDEF	Baseline: HD group significantly worse in recognition of anger, fear, and sadness. Follow-up: No between-groups or within-groups analysis available.
[43]	Mexico	Between-groups	Manifest	12 42.7 (median) 67	Relatives: 12 44.7 (median) 58 Manifest Ctrl: 12	Visual Auditory	Eyes Faces	POFA RMET	HD group significantly worse on emotion recognition through eyes (RMET) but not faces (POFA).

					37.1 (median) 75				
44	German UK	Case series	Manifest	<u>Case 1</u> 1 41 0 <u>Case 2</u>: 1 30 0	15 43.3 (12.1) 53	Visual	Faces	EHT	Significant impairment of recognition of disgust and fear in both patients. Normal recognition of happiness, surprise, sadness, and anger.
45	UK	Between-groups	Premanifest Manifest	<u>Premanifest:</u> 20 47.6 (8.45) 40 <u>Manifest:</u> 20 38.4 (9.5) 65	20 47.9 (9.3) 40	Visual	Faces	POFA	Manifest impaired on anger, fear, disgust, and sadness compared to both controls and premanifest; no significant differences on other emotions. Premanifest not significantly different from controls on any emotion. Benton, phonemic fluency, semantic fluency, and WAIS-III vocabulary found to contribute to explanation of variance in emotion recognition in regression models.
46	France	Single case study	Manifest	1 47 (NR) 0	20 46 (7.5) NR	Visual Multimodality	Eyes Faces	ADFES RMET	No significant impairment observed.
47	Australia	Between-groups	Manifest	17 61 (12) 47	24 62 (9) 63	Multimodality	Video Vignettes	TASIT-EET	HD group significantly worse than controls on negative emotions (anger, fear, disgust, sadness), but not positive ones (happiness, surprise) or neutral. No specific scores available for each emotion.

[48]	UK	Between-groups	Manifest	13 53.46 (5.11) 69	12 52.17 (7.907) 58.3	Visual	Faces Body language	BESST	HD group significantly worse than controls in recognising disgust and anger from facial stimuli, and fear, sadness, and neutral stimuli from body language.
[49]	Australia	Between-groups	Manifest	14 54.6 (11.17) 42.8	14 51.8 (8.37) 50	Visual Auditory	Faces Visual Vignettes Nonverbal Sounds	EHT FEEST Custom	HD group significantly worse than controls at recognising anger, disgust, fear, sadness and surprise from faces, but not happiness. Only disgust impaired from visual vignettes.
[50]	UK	Between-groups	Manifest	16 57.5 50	16 56.6 56.25	Visual	Eyes	RMET	HD group significantly worse than controls.
[51]	UK	Between-groups	Manifest	<u>Study 1</u>: 21 50.43 (8.70) 42.8 <u>Study 2</u>: 19 42.43 (11.35) 47.7	<u>Study 1</u>: Different for each task <u>Study 2</u>: 14 42.43 (11.35) 64	Visual Auditory	Faces Nonverbal Sounds Written Vignettes	POFA EHT Custom	HD group significantly worse at recognising anger, fear, and disgust in facial and auditory tasks. Only anger impaired on written vignettes.
[52]	Denmark	Between-groups	Premanifest Manifest	<u>Premanifest</u>: 50 37 (NR) 42 <u>Manifest</u>: 50 51 (NR) 40	39 41 (NR) 56	Visual	Faces Eyes Video Vignettes	EHT RMET TASIT-EET	No significant impairments observed in the Premanifest HD group across all tasks compared to controls. Manifest HD group significantly worse than controls at recognising emotions through faces (EHT), eyes (RMET), and video vignettes (TASIT-EE).

[53]	Denmark	Between-groups (longitud.)	Premanifest Manifest	T1 Premanifest: 50 36.5 (8.8) 42 T1 manifest: 48 51.2 (12) 39.5 T2 premanifest: 34 41.15 (8.5) 38.2 T2 manifest: 46 52.89 (11.6) 45.6	46 42.0 (13.4) 56.5	Visual	Faces Eyes	EHT RMET POFA	Premanifest HD group not significantly impaired compared to controls at either baseline or 6-year follow up. Manifest HD group consistently impaired compared to premanifest HD group and controls at both baseline and 6-year follow-up.
[54]	UK	Between-groups	Premanifest Manifest	Premanifest: 21 37.2 (7.9) 52 Manifest: 40 48.5 (9.6) 50	20 44.9 (10.5) 65	Visual	Faces	POFA	HD group significantly worse than controls at recognising, surprise, disgust, anger and fear, and worse than premanifest HD at recognising disgust and anger.
[55]	UK	Between-groups	Premanifest	11 NR	17 50.7 (14.3)	Visual Auditory	Faces	FEEST Custom	HD group significantly worse than controls at recognising anger, fear, disgust, sadness,

				NR	47		Nonverbal Sounds		and surprise, but not happiness, from facial stimuli. Impairment of fear, disgust, happiness, and surprise, but not anger and sadness from auditory stimuli.
56	Germany	Between-groups	Manifest	41 48.7 (10.0) NR	26 47.0 (9.5) NR	Visual	Faces Eyes	POFA RMET	HD group significantly worse than controls at recognising anger, fear, disgust, sadness, and surprise, but not happiness, from facial stimuli. Effect sizes highest for disgust and anger. General impairment in recognising emotions from eyes.
57	UK	Between-groups	Manifest	13 53.1 38.5	12 53.1 50	Visual	Visual Vignettes Written Vignettes	IAPS Custom	HD group significantly worse at recognising fear from visual vignettes but not disgust or happiness. No impairment on fear, disgust, or happiness with written vignettes.
58	Spain	Between-groups	Manifest	22 58.09 (9.73) 59	19 52.00 (9.69) 53	Visual	Eyes Faces	RMET POFA	HD group significantly worse than controls with both eyes (RMET) and faces (POFA).
63	France	Between-groups	Manifest	13 54.1 (7.2) 46	18 52.3 (5.4) 33	Visual	Faces	POFA KDEF	HD group significantly worse than control in recognition of fear, disgust, and sadness, but not fear, happiness and surprise.
64	France	Between-groups	Manifest	28 50 (8) 42.8	24 49 (10) 50	Visual	Faces	POFA KDEF	HD group significantly worse than control in recognition of anger, fear, disgust, happiness, sadness, and surprise.
75	Denmark	Between-groups	Premanifest Manifest	Premanifest: 40 41.3 (11.0) 40	32 48.1 (14.1) 59	Visual	Faces Eyes	EHT RMET	HD group significantly worse than controls across both measures.

				<u>Manifest</u>: 40 51.7 (11.9) 47.5					
76	USA	Between-groups	Premanifest	21 54 (9) 57	16 56 (12) 62	Visual	Faces	ERT	Premanifest HD group significantly less likely than controls to recognise anger, fear, and sadness, but not disgust, happiness, surprise, or neutral stimuli.
77	UK	Between-groups	Manifest	10 47 (9) 50	12 57 (9) 33	Visual Auditory	Faces Eyes Nonverbal Sounds	POFA FEEST MFS Custom	HD group significantly impaired on both visual and auditory tasks, with predominant impairment for negative emotions.
78	UK	Between-groups	Premanifest Manifest	<u>Premanifest</u>: 16 42.13 (13.49) NR <u>Manifest</u>: 16 13.25 (2.11) NR	28 46.7 (13.4) 54	Visual	Eyes	RMET	Both premanifest and manifest HD groups both significantly worse than controls. Manifest HD group significantly worse than premanifest HD group.
79	UK	Between-groups	Premanifest Early manifest Moderate manifest Late manifest	<u>Premanifest</u>: 29 43.5 (9.5) 51.7 <u>Early manifest</u>: 12 54.1 (11.5)	26 59 (11.7) 46.1	Visual	Eyes	RMET	No significant impairments observed in the Premanifest HD group compared to controls. All Manifest HD groups significantly worse than controls.

				25 Moderate manifest: 18 52.8 (14.4) 50 Late manifest: 20 56.1 (10.2) 35					
80	Australia	Between-groups	Manifest	11 56.82 (9.81) 37.5	11 55.64 (7.06) NR	Visual	Faces	KDEF	HD group significantly worse than controls with neutral, angry, and disgust facial expressions.
81	Austria	Between-groups	Manifest	18 51.9 (10.4) 44	18 49.2 (10.3) 44	Visual	Faces	KDEFS	HD group significantly less accurate than controls at recognising sadness, anger, and disgust, but not fear and surprise. HD group significantly better than controls at recognising happiness.
82	Portugal	Between-groups	Premanifest Manifest	Premanifest: 16 36.2 (1.8) 87.5 Manifest: 9 48.8 (4.6) 0	22 41.0 (2.3) 59	Visual	Faces	FEEST POFA	Premanifest HD group not significantly impaired compared to controls. Manifest HD group significantly impaired on all emotional stimuli compared to controls. Smaller impairment for happiness.
84	UK	Between-groups	Premanifest	20 45.0 (14.0)	26 45.7 (14.4)	Visual	Eyes	RMET	HD group significantly worse than controls.

				70	69.2				
85	UK	Between-groups	Premanifest	23 38.53 (11.24) NR	15 38.26 (11.82) NR	Visual	Faces	POFA	Premanifest significantly poorer at recognising disgust than controls; no difference on any other emotions.
86	Austria	Between-groups	Manifest	28 48.4 (9.4) 39	28 47.2 (7.5) 39	Visual	Faces	KDEFS	HD group significantly less accurate than controls at recognising anger, disgust, surprise, and sadness.
87	Austria	Between-groups	Manifest	18 51.9 (10.4) 44.4	18 49.2 (10.3) 44.4	Visual	Faces	KDEF	HD group significantly worse than controls for anger, but not fear, disgust, happiness, sadness, or surprise.
88	Germany	Between-groups	Premanifest	9 37.4 (5.4) 44	9 NR 44	Visual	Faces	FEEST	Premanifest HD group significantly worse than controls at recognising disgust, but not anger, fear, happiness, sadness, or surprise.
89	UK	Between-groups	Premanifest	16 43.81 (8.30) 75	14 39.43 (11.40) 71	Visual	Faces	POFA	No significant impairment observed.
104	France	Between-groups	Manifest	18 50.7 (8.8) 44.5	18 47.5 38.9	Visual	Eyes	RMET	HD group significantly worse than control on RMET.
105	Australia	Between-groups	Premanifest	17 43.8 (10) 47	13 42.0 (11.4) 30.7	Visual	Faces	EHT	No significant differences observed.
106	Australia	Between-groups	Premanifest Manifest	23 48.83 (8.90) 56	25 49.64 (8.86) 60	Visual	Faces	ADFES	HD group significantly worse at recognising happiness, anger, disgust sadness, and surprise.

[107]	Netherlands	Between-groups	Manifest	8 46.4 (11.2) 37.5	30 39 (11.1) 53	Visual	Faces	Custom	HD group impaired in recognition of disgust and anger, but not fear, happiness, sadness, and surprise.
[108]	Italy	Between-groups	Premanifest	18 35.6 (7.2) 50	18 37.3 (9.6) 50	Visual	Eyes	RMET	Premanifest HD group significantly worse than controls.
[109]	USA	Between-groups	Premanifest Manifest	Premanifest: 14 47.43 (10.83) 50 Manifest: 62 50.29 (13.12) 55		Visual	Faces	ACS-AN	Manifest HD group significantly worse than Premanifest HD group.
[110]	Spain	Between groups	Manifest	21 58.1 (9.7) 59.1	22 52 (9.7) 53	Visual	Faces	POFA	HD group significantly worse than controls.
[111]	Denmark	Between-groups	Manifest	52 51.0 (11.8) 40.3	166 47.9 (20.9) 58.4	Visual	Faces	EHT	HD group significantly worse than controls.

Note. ACS-AN = Advanced Clinical Solutions Affect Naming; ADEFS = Amsterdam Dynamic Facial Expression Set; EHT = Emotion Hexagon Test; ER = emotion recognition; ERT = Emotion Recognition Task; FEEST= Facial Expressions of Emotions: Stimuli and Tests (POFA + EHT); IAPS = Affective Picture System; JeFEE= Jerusalem Facial Expressions of Emotion; KDEF = Karolinska Directed Emotional Faces; MFS = Manchester Face Set; NimStim = NimStim Set of Facial Expressions; POFA= Pictures of Facial Affect; RMET = Reading the Mind in the Eyes Test; T/C = total or composite score.

Figure 1

PRISMA Flow Diagram for Selection of Studies

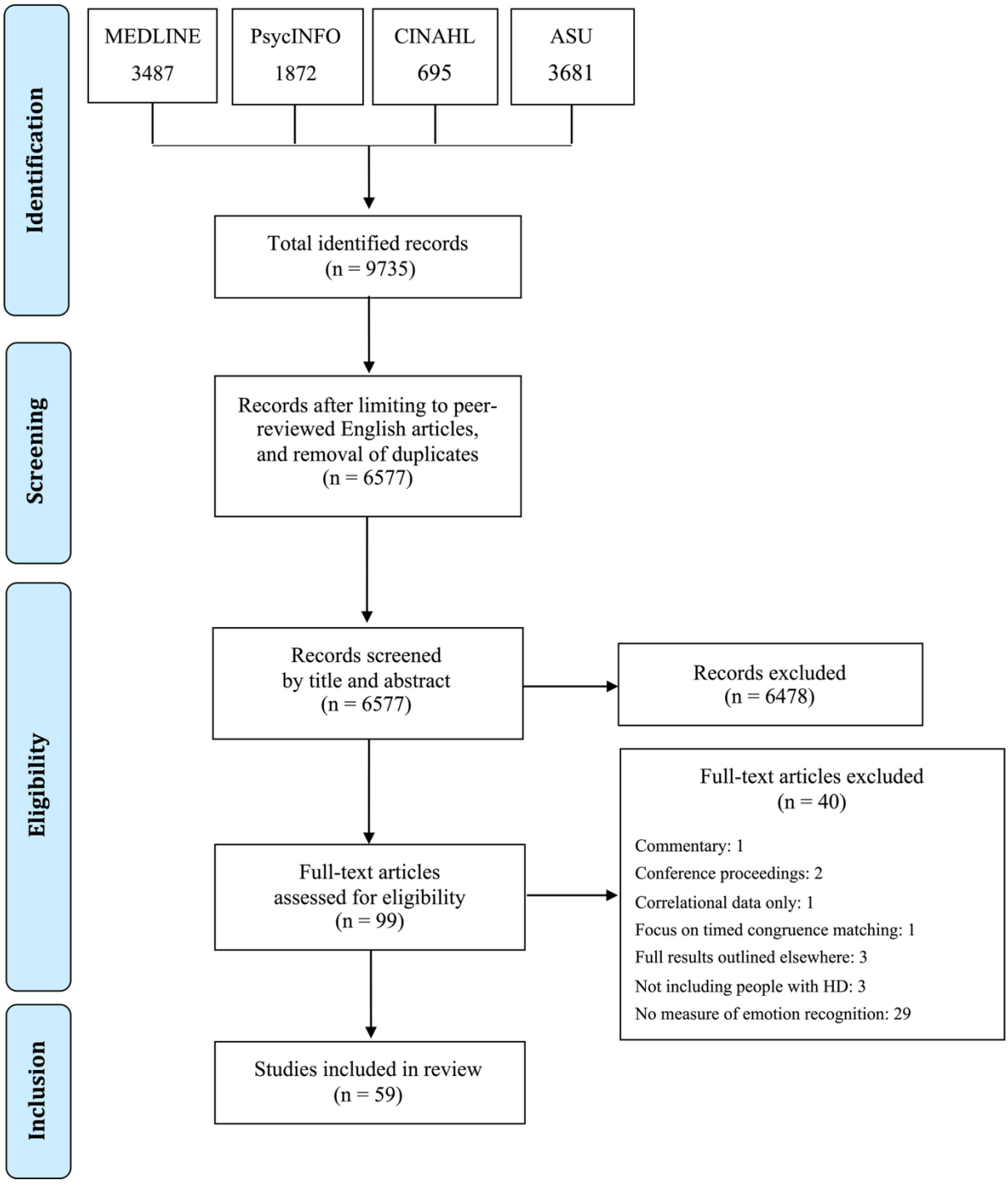
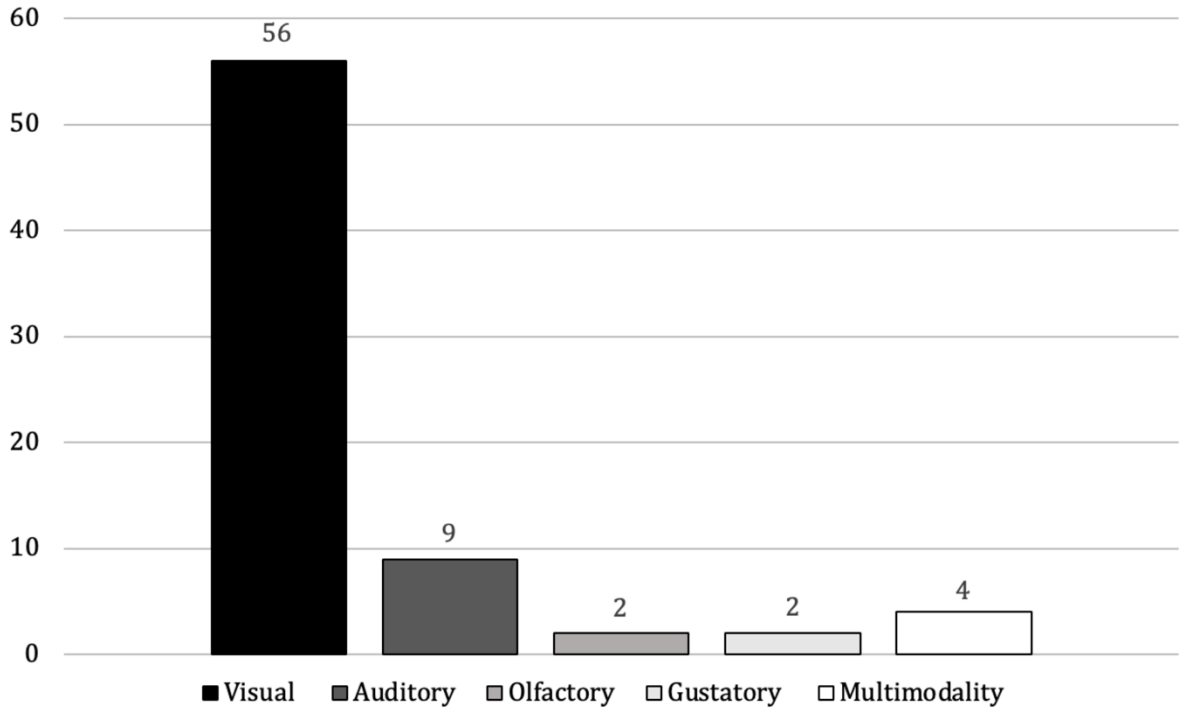


Figure 2

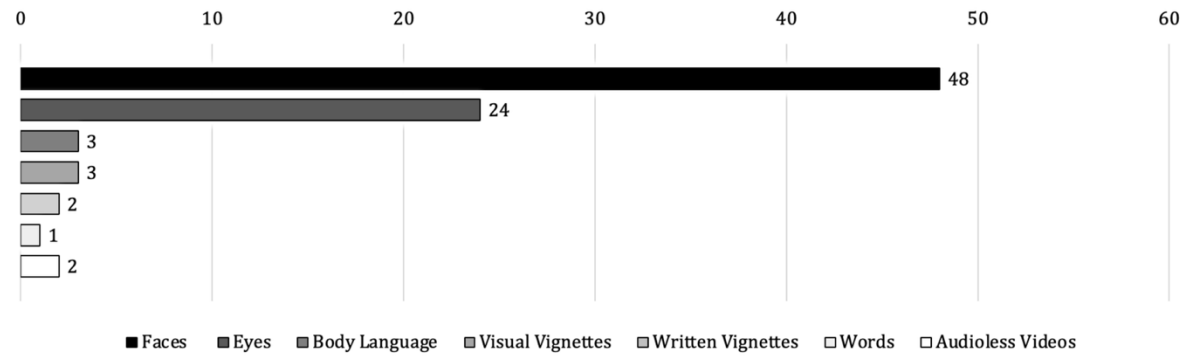
Emotion Recognition Modalities Across Included Studies



Note. Total included studies = 59. Some studies investigated multiple modalities.

Figure 3

Types of Visual Stimuli Across Included Studies



Note. Total visual studies = 56. Some studies adopted multiple stimulus types.