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REVIEW ARTICLE



# Psychosocial functioning in patients with altered facial expression: a scoping review in five neurological diseases

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**ABSTRACT**

**Purpose:** To perform a scoping review to investigate the psychosocial impact of having an altered facial expression in five neurological diseases.

**Methods:** A systematic literature search was performed. Studies were on Bell's palsy, facioscapulohumeral muscular dystrophy (FSHD), Moebius syndrome, myotonic dystrophy type 1, or Parkinson's disease patients; had a focus on altered facial expression; and had any form of psychosocial outcome measure. Data extraction focused on psychosocial outcomes.

**Results:** Bell's palsy, myotonic dystrophy type 1, and Parkinson's disease patients more often experienced some degree of psychosocial distress than healthy controls. In FSHD, facial weakness negatively influenced communication and was experienced as a burden. The psychosocial distress applied especially to women (Bell's palsy and Parkinson's disease), and patients with more severely altered facial expression (Bell's palsy), but not for Moebius syndrome patients. Furthermore, Parkinson's disease patients with more pronounced hypomimia were perceived more negatively by observers. Various strategies were reported to compensate for altered facial expression.

**Conclusions:** This review showed that patients with altered facial expression in four of five included neurological diseases had reduced psychosocial functioning. Future research recommendations include studies on observers' judgements of patients during social interactions and on the effectiveness of compensation strategies in enhancing psychosocial functioning.

**> IMPLICATIONS FOR REHABILITATION**

- Negative effects of altered facial expression on psychosocial functioning are common and more abundant in women and in more severely affected patients with various neurological disorders.
- Health care professionals should be alert to psychosocial distress in patients with altered facial expression.
- Learning of compensatory strategies could be a beneficial therapy for patients with psychosocial distress due to an altered facial expression.

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Bell's palsy; facioscapulohumeral muscular dystrophy; myotonic dystrophy; Moebius syndrome; Parkinson's disease; psychosocial functioning; facial expression; facial weakness

## Introduction

Facial expressions are important in conveying emotions, in identifying communicative intentions in social interactions and are part of a sophisticated communication system [1,2]. Within seconds, people form their first impression based on someone's facial appearance [3]. People, especially women, with more pronounced positive facial expressions are perceived more positively in first impressions [4]. Besides expressing emotions through facial expression, understanding and sharing expressed emotions of others by mimicking their facial expression is important in social bonding [5]. Moreover, facial signals also pay an important role in communication meaning in social interaction [6,7]. When facial expression is impaired others could misinterpret non-verbal communication, which could implicate a possible mismatch between experienced emotions or communicative intentions and the information conveyed by the face.

Observers' judgements of patients with facial palsy have been investigated in various studies. In contrast, the patients' perspective on psychosocial functioning related to altered facial expression is less investigated. Patients with facial palsy are perceived

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by observers as less trustworthy, more distressed, and less attractive [8,9]. Observers classify these patients as displaying a negative effect, even when they are smiling [10]. Judgements by observers are more negative than patients' own perceptions on attractiveness and quality of life [11]. People who are not fully capable of conveying emotions and meaning through facial expressions can therefore experience impairments in social interactions.

During outpatient contacts, we noticed that facioscapulohumeral muscular dystrophy (FSHD) patients frequently addressed the experienced difficulties of having an altered facial expression because of facial weakness. This is illustrated by the patient with FSHD and reduced facial expressions in Figure 1. The experienced difficulties were the reason to conduct a literature study on the psychosocial impact of having an altered facial expression. In this scoping review, we aim to investigate the psychosocial consequences of having an altered facial expression in different neurological diseases. A systematic review on psychosocial distress in facial palsy patients has recently been performed, but other neurological diseases were not included [12]. Our review has a different starting point by including five neurological disorders (i.e., neuromuscular and movement disorders) and also including smaller studies (case reports) to maximize existing knowledge to give insight on the common themes of having an altered facial expression on psychosocial functioning.

We included five neurological diseases: Bell's palsy, FSHD, Moebius syndrome, myotonic dystrophy type 1, and Parkinson's


**Figure 1.** Facial expressions of a FSHD patient with facial weakness.

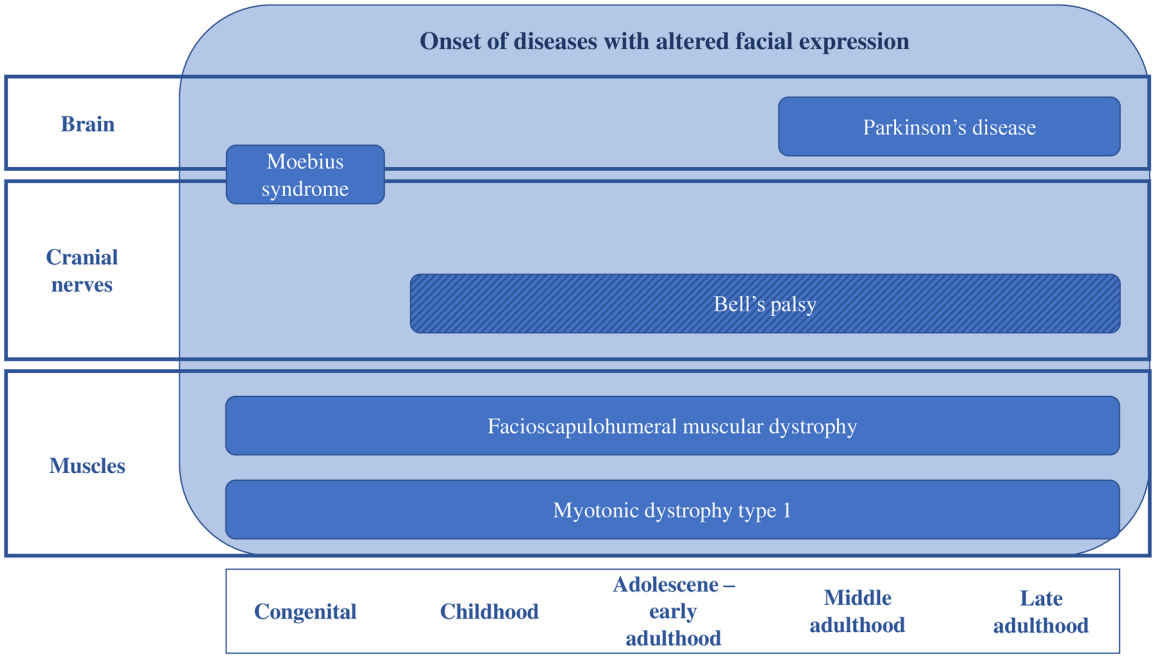


**Figure 2.** Overview of time of onset and origin of pathophysiology of altered facial expression in the included diseases. *X*-axis: time of disease onset. *Y*-axis: anatomical localisation of pathophysiology. Disease onset: diagonal stripes represent an acute onset; equal colour represents a gradual onset. *Bell's palsy* is an idiopathic unilateral peripheral facial neuropathy with acute onset. Pathogenesis is unclear, but auto-immune response, inflammation, and nerve compression are possibly part of it [68]. *Facioscapulohumeral muscular dystrophy* is a slowly progressive muscle disease, mostly due to a repeat contraction of D4Z4 macrosatellite array on chromosome 4. Facial weakness is one of the symptoms and occurs often early in disease progression [69]. *Moebius syndrome* is a congenital disease with unilateral or bilateral facial palsy and abducens nerve palsy, due to cranial nerve nuclei impairment. Impairment of cranial nerve eight (cochlear branch) and dysmorphisms are also described in Moebius syndrome [45]. It is a rare disease with a prevalence of approximately 1/50 000 [70]. *Myotonic dystrophy type 1* is a muscle disease caused by an CTG triplet expansion in the DMPK gene. Facial weakness is one of the symptoms and leads to a disease specific facial appearance with tented upper lip. Other symptoms include myotonia and muscle weakness in distal limbs, and trunk muscles, cardiac conduction defects, respiratory insufficiency, and cataract [71]. *Parkinson's disease* is a neurodegenerative disease due to loss of dopaminergic neurons in substantia nigra, which causes dopamine depletion. It consists of four cardinal symptoms: bradykinesia, postural instability, rigidity, and resting tremor. One of the manifestations of bradykinesia is hypomimia, what can cause an altered facial expression [72].

disease. These diseases have a very different pathophysiology, but can all cause an altered facial expression. This selection of diseases was chosen carefully to ensure a diverse overview of possible causes of altered facial expression: acute versus gradual onset, congenital versus acquired disease, and different aetiologies. Figure 2 highlights the similarities and differences between the diseases. The psychosocial study outcomes in these diseases are expected to create an understanding of the possible psychosocial impairments of having an altered facial expression and to help identify future research directions for improving verbal and non-verbal communication, and thus psychosocial functioning.

## Materials and methods

A literature search was conducted to get an overview of published literature on psychosocial functioning in five neurological diseases with altered facial expression.

### *Protocol*

A study protocol was registered in advance at CRD42020203220.

### *Type of study*

Quantitative, qualitative, and mixed methods studies were all eligible. Case reports were also included, because we expected literature to be scarce and wanted to give a complete overview of the available peer reviewed literature. Studies had to be published in English or Dutch. Conference abstracts were excluded.

### *Participants*

Participants in the included studies must have one of the following diseases: Bell's palsy, FSHD, Moebius syndrome, myotonic dystrophy type 1, or Parkinson's disease. We excluded other causes of facial paralysis. Since we expected studies in Bell's palsy to also consist of other acquired causes of facial paralysis, we only included studies in which the majority (≥50%) of participants was diagnosed with Bell's palsy. By doing so, we attempted to maximize the inclusion of potentially relevant studies. If other aetiologies of facial palsy were included, percentages per aetiology were described in Table 1. Studies including myotonic dystrophy were only included when patient population consisted of myotonic dystrophy type 1, because facial weakness is only a prominent symptom in myotonic dystrophy type 1 and not in type 2. Studies with Parkinson's disease were only included when the influence of hypomimia on psychosocial aspects was investigated.

### *Outcomes measures*

Quantitative articles were included when the outcome measurements of the study related to psychosocial functioning. Qualitative articles were included if psychosocial functioning was part of the study design in the interview guide or in the focus group guide. Psychosocial functioning was defined as emotional and social well-being and therefore included communication, participation, behaviour, quality of life, occupation, emotional state, or satisfaction. In addition to these subjects, there was also focus on swallowing problems, since this can negatively impact psychosocial functioning (embarrassment, anxiety, and avoiding eating in company) [41]. Studies focussing on swallowing

**Table 1.** Overview of included articles.

| First author Year Country | Study design Level of health care | Participants: Sample size (% male) | Participants: Mean age (years) | Participants: Time since diagnosis | Assessment tools psychosocial functioning | Methods | Psychosocial outcomes |
|---|---|---|---|---|---|---|---|
| *Bell's palsy* | | | | | | | |
| Bylund [13] 2020 SWE | Prospective cohort study THC | 96 (63%) | 49 (SD = 17) | *m* = 7 d (SD = 8) | FaCE Scale FDI | Facial grading system: Sunnybrook Facial Grading Score and HB-Score. Visit 1: day of onset Bell's palsy Visit 2: mean day 53 (SD = 40) Visit 3: mean day 137 (SD = 105) Visit 4: mean day 260 (SD = 140) | FaCE Scale (social function): visit 1: 69; visit 2: 85; visit 3: 81; visit 4: 74. Visit 1: fair correlation with Sunnybrook for men and women. Visits 2–4: higher correlation with Sunnybrook Score for women. FDI social/well-being function: visit 1: 75; visit 2: 79; visit 3: 80; visit 4: 79 FaCE domain social function and FDI social/well-being function at visits 1–3: women scored lower; Sunnybrook Scores: similar |
| Huang [14] 2012 CHN | Case-control study THC | Patients: 306 (58%) Controls: 320 (50%) | 37.5 (SD = 12.2) | 1–3 d: 53% 3–7 d: 47% | K10 FDI | Facial grading system: HB-Score Severity of disease: mild (HB-Score 1–3); severe (HB-Score 4–6) Onset time: acute (within 3 d) and sub-acute (3–7 d) | K10: controls: 13.43. Patients: 27.09* Patients: women: 28.07. Men: 26.38* Correlation with severity facial palsy: mild: 24.22. Severe: 31.49* Subacute patients: 28.46. Acute patients: 26.24* FDI social life function: 66.52 |
| Kim [15] 2018 USA | Questionnaire study PHC | 160 (9%) | 45.1 (SD = 12.6) | <1 y: 12% 1–2 y: 9% 2–3 y: 17% >3 y: 63% | CPIB Short Form FaCE Scale | Only unilateral facial palsy. 25 participants had prior treatment. Bell's palsy: 54% Tumour: 26% Other causes: 21% | CPIB Short Form: 0.16 (0.88) logits. No effect for age, prior treatment, cause, and duration of disease. Correlation CPIB Short Form and FaCE Scale: *r* = 0.47* FaCE Scale: Bell's palsy vs. tumour: better scores: facial movement subscale; worse scores: eye comfort and facial comfort subscales. |
| Krane [16] 2020 USA | Cross-sectional study THC | 56 (30%) | 56.4 (SD = 15.5) | *m* = 11.9 y (SD = 13.3) | BFNE-II SAQ-A30 SAD Scale SF-6D FaCE Scale | Level of facial impairment: self-perceived Bell's palsy: 63% Neoplastic: 21% Other causes: 17% | Moderate/severe impairment: higher scores on psychosocial impairment surveys (compared with none or mild impairment): SAQ: *r* = 47.9 vs. *r* = 31.5*, SAQ-A30: *r* = 90.3 vs. *r* = 75.2*, SAD: *r* = 14.2 vs. *r* = 7.9*, BFNE-II: *r* = 35.2 vs. *r* = 27.6*. And lower score on SF-6D: *r* = 0.69 vs. *r* = 0.78*. FaCE total score: women: 52.5, men: 75.7*; idiopathic/infectious: 66.13, neoplastic/traumatic/iatrogenic: 44.78* |
| Lee [17] 2019 KOR | Cohort study PHC | Part 1: 17.630 (48%) Part 2: 305.340 (34%) | Part 1: 20–39: 25% 40–59: 44% >59: 31% Part 2: 20–39: 32% 40–59: 39% >59: 29% | Part 1: date to depression: controls: *m* = 43 mth BP: *m* = 37 mth Part 2: date to BP: controls: *m* = 53 mth depression: *m* = 50 mth | None | Data were used from the Korean National Health Insurance Service-National Sample Cohort. All citizens from South Korea are part of this database. Part 1: Bell's palsy patients and controls were matched. Control group had not been diagnosed with Bell's palsy in 2002–2013. Part 2: Patients with depression were matched with controls. Control group had never been diagnosed with depression in 2002–2013. | Part 1: depression rate Bell's palsy: 5.8%. Controls: 4.1%* Women ≥40 y: increased risk of developing depression. Hazard ratio 40–59 y: 1.45* and ≥60 years: 1.50*. Part 2: rate of Bell's palsy was similar in depression group and control group: 0.4%*. |

(*Continued*)

**Table 1.** Continued.

| First author<br>Year<br>Country | Study design<br>Level of health care | Participants<br>Sample size (% male) | Mean age (years) | Time since diagnosis | Assessment tools psychosocial functioning | Methods | Psychosocial outcomes |
|---|---|---|---|---|---|---|---|
| Pouwels<br>[18]<br>2016<br>NLD | Cohort study<br>THC | Patients: 59 (37%)<br>Controls: 59 (34%) | Patients:<br>56 (±15)<br>Controls: 40 (±16) | *m* = 5.4 y (SD = 6.1) | HADS | Bell's palsy: 51%<br>Herpes Zoster: 17%<br>Acoustic neurinoma: 12%<br>Other cause: 20% | Mild anxiety: facial palsy 20.3%, controls: 6.8%*. No significant difference in moderate or severe anxiety.<br>Mild depression: facial palsy: 13.6%, controls: 3.4%*<br>Moderate depression: facial palsy: 11.9%, controls: 0%*<br>No significant difference in severe depression rate. |
| Smith<br>[19]<br>1994<br>GBR | Questionnaire study<br>PHC | 120 (23%) | Men: 46 (18–72)<br>Women: 52 (16–83) | Men: *m* = 13.7 y<br>Women:<br>*m* = 11.5 y | NVQ | Bell's palsy: 75%<br>Other cause: 25%<br>Patients were recruited, while waiting for attending in an ENT clinic. | 30% of patients with residual facial weakness did not noticed effect in social situations.<br>45% noticed a small effect and 25% noticed a profound effect. |
| Weir<br>[20]<br>1995<br>GBR | Questionnaire study and qualitative study<br>SHC | 20 (40%) | 41 (15–78) | *m* = 65 d<br>(6 d–7 y) | FSI<br>GHQ<br>FDQ | Severity of facial palsy:<br>International Facial Palsy Grading:<br>Median: III<br>Semi-structured interview was also conducted. | FSI: dissatisfaction with facial feature(s): 16 patients<br>GHQ: clinical disability for anxiety and depression: 5 patients<br>FDQ: most affected category: social activities. No affected functionality: 5 patients.<br>Interview: changes in attitude of others towards them: 12 patients. Negative experience: 6 patients. Positive experience: 4 patients. Mixed experience: 2 patients. 8 patients felt less attractive to others.<br>Low association between severity of facial palsy and change in psychosocial functioning. |
| *Facioscapulohumeral muscular dystrophy* | | | | | | | |
| Bakker<br>[21]<br>2017<br>NLD | Qualitative study<br>SHC and THC | 25 (56%) | 56 (range 24–77) | *m* = 16 y (range 1–49 y) | None | All FSHD-patients were under the care of a rehabilitation specialist.<br>The semi-structured interview consists of personal experiences, possible difficulties, and social environment. | Acceptance of illness is difficult, because of the progression of limitations. Facial weakness has a major impact, because of the visibility in mirrors and pictures and has a negative effect on non-verbal communication. Diagnosis often led to more understanding of others and less negative feelings. |
| Hamel<br>[22]<br>2019<br>USA | Cross-sectional study<br>SHC and THC | 328 (46%) | 55 (SD = 13.4) | 0–10 y (*n* = 28)<br>11–20 y (*n* = 56)<br>21–30 y (*n* = 65)<br>31–40 y (*n* = 71)<br>≥41 y (*n* = 84) | Life impact score | All patients were registered in the National Registry of Myotonic Dystrophy and Facioscapulohumeral Muscular Dystrophy Patients and Family Members.<br>This survey consists of 15 symptomatic themes and 274 symptoms. Patients had to state how much a symptom impacts their life. | Decreased performance and satisfaction in social situations: respectively 77% and 72%. Communication difficulties: 33%.<br>Employed vs. unemployed: communication difficulties 47% vs. 24%*, decreased performance in social situations: 86% vs. 69%*<br>Longer duration of disease: higher prevalence of decreased performance in social situations.<br>No differences between men and women in theme prevalence.<br>A higher life impact score in decreased performance in social situations was observed in women compared with men: 1.84 vs. 1.48*. |

(*Continued*)

**Table 1.** Continued.

| First author<br>Year<br>Country | Study design<br>Level of health care | Participants<br>Sample size (% male) | Mean age (years) | Time since diagnosis | Assessment tools psychosocial functioning | Methods | Psychosocial outcomes |
|---|---|---|---|---|---|---|---|
| Johnson<br>[23]<br>2012<br>USA | Qualitative study<br>THC | 20 (35%) | 51 (SD = 12.1) | Not reported | None | In-depth interviews were conducted for identification of symptoms with the most impact on quality of life and how patients are affected by FSHD.<br>251 symptoms and 1.375 quotes were collected. | Social role limitations were mentioned more than facioscapulohumeral weakness, when patients were asked what affected them most in daily life.<br>Impaired facial expression, inability to smile and facial weakness were mentioned by 5 patients.<br>Dysphagia was mentioned by 3 patients.<br>Inability to make facial expression was mentioned by 2 patients. |
| Mul<br>[24]<br>2019<br>USA | Quantitative study<br>THC | Patients: 43 (56%)<br>Controls: 35 (49%) | Patients: 53 (SD = 13.1)<br>Controls: 40 (SD = 13.2) | *m* = 30 y (SD = 15.2) | SWAL-QOL<br>CPIB | Iowa Oral Performance Instrument was used for obtaining strength and endurance measurements of lip and cheek compression and tongue elevation. | Cheek compression strength was reduced in FSHD patients compared with controls. Lip compression strength was not reduced, and tongue elevation strength was only reduced in men.<br>SWAL-QOL: scores were lower on all domains compared with normative values.<br>CPIB: 42% of FSHD patients had no communication difficulties. 35% of FSHD patients scored lower than 75% of the maximum score, indicating communication difficulties. Lowest scores: say something quickly and get a turn in fast moving conversations (both in 47%).<br>Correlation SWAL-QOL scores and cheek compression strength: $r$ = 0.382*. Correlation CPIB scores and cheek compression strength: $r$ = 0.398*. |
| *Moebius syndrome* | | | | | | | |
| Bogart<br>[25]<br>2010<br>USA | Questionnaire study<br>Unknown | Patients: 37 (38%)<br>Controls: 37 (38%) | Patients: 38 (SD = 13.7)<br>Controls: 35 (SD = 12.5) | Not applicable (congenital disease) | HADS<br>SWLS<br>TSBI<br>FECQ | Patients were recruited with help of Moebius Syndrome Foundation.<br>37 controls were selected from 249 participants to match age and gender with Moebius patients.<br>FECQ was created for this study. | HADS: anxiety: patients: 7.9; controls: 7.3. Depression: patients: 4.0; controls: 4.4<br>SWLS: patients: 21.6; controls: 22.7<br>TSBI: patients: 36.4*; controls: 40.2*<br>FECQ score: patients: 16.2*; controls: 25.9*<br>FECQ score correlations: not statistically significant with depression, anxiety, social competence, or satisfaction with life. |
| Bogart<br>[26]<br>2012<br>USA | Qualitative study (focus group)<br>Unknown | 12 (50%) | 41 (18–60) | Not applicable (congenital disease) | None | Two moderators (both diagnosed with Moebius syndrome) led a discussion, with 7 open-ended questions. Discussion took place during the Moebius Syndrome Foundation Conference. | Social (dis)engagement: plays a prominent role. Others do not know how to react. Compensation of facial weakness with tone of voice, humour, clothing, verbal disclosure, and speaking clearly.<br>Resilience: some patients had low confidence and others displayed confidence and persistence.<br>Being (mis)understood: barriers: lack of facial expression, speech difficulties, others not knowing the disease and stigma. Familiar people (family/friends) see the person behind Moebius. |

(*Continued*)

**Table 1.** Continued.

| First author<br>Year<br>Country | Study design<br>Level of health care | Participants<br>Sample size (% male) | Participants<br>Mean age (years) | Participants<br>Time since diagnosis | Assessment tools psychosocial functioning | Methods | Psychosocial outcomes |
|---|---|---|---|---|---|---|---|
| Bogart<br>[27]<br>2015<br>USA | Qualitative study (focus group)<br>Unknown | 10 (30%) | 14.3 (12–17) | Not applicable (congenital disease) | None | Two psychologists (one diagnosed with Moebius syndrome) led a discussion, with 8 open-ended questions. This was during the Moebius Syndrome Foundation Conference. | Subjects: fear of being misunderstood, insecure to start a conversation, prejudice, and social exclusion.<br>Compensating lack of facial expression by using compensatory expressions: tone of voice and body language. Positive reappraisal on negative experiences in social interactions. |
| Briegel<br>[28]<br>2011<br>DEU | Questionnaire study<br>Unknown | 17 (53%) | 11.6 (9–15) | Not applicable (congenital disease) | AFS<br>DIK<br>PFK 9–14<br>SDQ-Deu | Patients filled in: AFS, DIK, PFK 9–14. Primary caregivers filled in: SDQ-Deu<br>*t*-scores of ≥63 were defined as clinical. | AFS and DIKJ: lower in patients compared with normative data.<br>PFK 9–14: self-perception of impulsivity was lower in patients compared with normative data.<br>SDQ: total score: one patient was rated clinical (5.9% vs. 10% in normative data). Social problems: 29.4% in patients vs. 10% in normative data and a correlation with age $r = 0.707$. Emotional problems: 17.6% in patients vs. 10% in normative data. |
| Strobel<br>[29]<br>2016<br>DEU | Questionnaire study<br>Unknown | 26 (58%) | 11.3 (4–17) | Not applicable (congenital disease) | KINDL<br>SDQ | Patients were recruited with help of the German Moebius Syndrome Association.<br>Patients (age: 11–17) and parents (one per family) filled in the questionnaires. | KINDL: parent ratings and self-reports were lower compared with normative data for subscale friends ($t = -3.71$). Total scores failed significance.<br>SDQ: parent ratings were higher for total difficulties ($t = 2.37$) and peer problems ($t = 3.54$) compared with normative data. These scores were higher than self-reports. |
| *Myotonic dystrophy type 1* | | | | | | | |
| Bungener<br>[30]<br>1998<br>FRA | Qualitative study<br>THC | MD: 15 (33%)<br>FSHD: 11 (36%)<br>Controls: 14 (36%) | MD: 36.8 (20–54)<br>FSHD: 33.2 (21–48)<br>Controls: 35.7 (20–55) | Not reported | MADRS<br>HDRS<br>COVI<br>TYR<br>AT<br>EHD<br>PAS<br>SAS | Participants were matched for age, sex, and education.<br>Semi-structured interviews last for 45 min. | Depression ratings (MADRS/HDRS): highest score in FSHD patients (only significant with controls).<br>Anxiety (COVI/TYR): no significant differences between groups.<br>Emotional deficits (part of EHD) and emotional blunting (AT): MD patients have highest scores.<br>Social anhedonia (SAS): no differences.<br>Physical anhedonia (PAS): MD and FSHD patients both higher than controls. |
| *Parkinson's disease* | | | | | | | |
| Gunnery<br>[31]<br>2016<br>USA | Cross-sectional study<br>THC | PD: 40 (68%)<br>Care partners: 40 (33%) | PD: 66 (SD = 7.4)<br>Care partners: 65 (SD = 7.1) | Not reported | SINHP<br>SSCI<br>MOS-SSS<br>GDS | Experienced hypomimia measurements: asking patients and partners about difficulties of showing facial expressions. | Patients with less facial expression: more social rejection ($r = 0.41$*) and unrelated to enjoyment of care partners.<br>Depression is correlated with facial expressivity ($r = 0.56$*) and social rejection ($r = 0.70$*).<br>Care partners who rated patients with less facial expressivity: reported more social rejection ($r = 0.35$*) and less partner enjoyment ($r = -0.55$*). |

(*Continued*)

**Table 1.** Continued.

| First author<br>Year<br>Country | Study design<br>Level of health care | Participants<br>Sample size (% male) | Participants<br>Mean age (years) | Participants<br>Time since diagnosis | Assessment tools psychosocial functioning | Methods | Psychosocial outcomes |
|---|---|---|---|---|---|---|---|
| Hemmesch<br>[32]<br>2009<br>USA | Qualitative observational study<br>THC | PD: 12 (50%)<br>Observers: 58 (31%) | PD: 65 (SD = 7.0)<br>Observers: 76 (SD = 8.1) | Not reported | PNSEA<br>PDQ-39<br>TIPI | Facial masking: 6 low and 6 high (assessed by 5 neurorehabilitation researchers).<br>PD-patients were filmed during an interview. Observers (age 55+) watched and rated clips (filtered for verbal content). Two questions were asked about likelihood of forming a relation with the PD patient.<br>Social support and strain were assessed with created instruments. Observer ratings were compared with self-reports. | Higher facial masking in women: less supportive (compared with women with low facial masking) and less desirable to form a relationship with (compared with men with similar facial masking). Self-reports did not differ in social supportiveness between low and higher facial masking.<br>Social supportive: In men, there was no difference.<br>Social strain: men with low facial masking were rated as more straining compared with men with high facial masking. In women, there was no difference. |
| Hemmesch<br>[33]<br>2014<br>USA | Qualitative observational study<br>THC | PD: 24 (50%)<br>Observers: 59 (41%) | PD: 69.1 (SD = 7.6)<br>Observers:<br>76.4 (SD = 6.5) | Not reported | PNSEA | Facial masking: 12 low and 12 high (assessed by trained raters).<br>Observers rated clips of interviews with PD-patients. Ratings involved: perceived supportiveness, caregiving expectations, and estimating reciprocity. Ratings were combined: social positivity score. Also interest in relationship with PD patients was rated. | High facial masking: less interest to form a relationship with ($r$ = 0.76*) and lower social positivity score ($r$ = 0.66*)<br>Men vs. women:<br>High facial masking: less interest to form a relationship with women ($r$ = 0.39*) and women were rated as less social positive ($r$ = 0.59*) |
| Kang<br>[34]<br>2019<br>KOR | Qualitative observational study<br>THC | PD: 20 (45%)<br>Controls:<br>20 (35%) | PD: 59 (SD = 8.6)<br>Controls: 59 (SD = 6.9) | Not reported | MWA<br>PDQ-39 | PD patients stopped with dopaminergic drugs 12 h before participation.<br>Clips were shown: observations afterwards for spontaneous and active mimicry. Muscle response (zygomaticus and corrugator muscle) was measured with EMG. | Patients vs. controls: reduced facial mimicry for emotional expressions in both spontaneous as active mimicry.<br>Mean and max. EMG activity of m. zygomaticus in spontaneous mimicry correlates with patients' well-being score, respectively, $r$ = 0.47* and $r$ = 0.48*. This correlation was weaker in voluntary mimicry: mean EMG activity ($r$ = 0.33*) and max. EMG activity: ($r$ = 0.35*). No correlation between m. corrugator activity and negative emotions. |
| Ma<br>[35]<br>2019<br>USA | Cross-sectional study<br>THC | 90 (62%) | 65 (SD = 9.7) | $m$ = 7.3 y (SD = 7.0) | SSCI<br>GDS<br>PDQ-39 | Facial masking severity was self-reported. Disease severity was measured with MDS-UPDRS. Patients had for the most part mild to moderate disease severity. | PDQ-39: quality of life was worse in women (mean = 30.74*) than men (mean = 24.30*). Positive correlation (stronger in women) between severity of facial masking and worse QoL with stigma as strong mediator.<br>Correlation between experienced stigma and QoL ($r$ = 0.82*). |

(*Continued*)

**Table 1.** Continued.

| First author<br>Year<br>Country | Study design<br>Level of health care | Participants<br>Sample size (% male) | Mean age (years) | Time since diagnosis | Assessment tools psychosocial functioning | Methods | Psychosocial outcomes |
|---|---|---|---|---|---|---|---|
| Pentland<br>[36]<br>1988<br>GBR | Qualitative observational study<br>Unknown | PD: 4 (100%)<br>IHD: 4 (100%) | PD: 53<br>IHD: 55 | PD: *m* = 3.3 y<br>IHD:<br>*m* = 3 y | NVQ | 19 students (3rd year of speech pathology and therapy) watched muted video clips and give their first impressions on: intellect, mood, and personality trait.<br>Control group: 4 patients with myocardial infarction in clinical history. | Observers:<br>PD patients: more anxious, angry, bored, sad, suspicious, and tense compared with IHD patients. Personality PD patients: more introvert, passive, dependent, and dissatisfied compared with IHD patients. |
| Schwartz<br>[37]<br>2017<br>CAN | Qualitative observational study<br>THC and SHC | *Part 1:*<br>PD: 17 (47%)<br>Controls: 20 (50%)<br>Raters: 10 (0%)<br>*Part 2:*<br>PD: 17 (47%)<br>Controls:<br>20 (50%) | *Part 1/2:*<br>PD: 64.7 (SD = 6.8)<br>Controls:<br>62.8 (SD = 7.4)<br>*Part 1:* Raters: 25.6 (SD = 2.5)<br>*Part 2:*<br>Raters: 67.0 (SD = 7.5) | *Part 1/2:*<br>PD: *m* = 9.9 y (SD = 6.2) | CPIB | *Part 1:* Spontaneous facial expressions of emotions in PD-patients. Controls and PD-patients watched videos which evoke emotions. Raters watched muted video clips of these responses. Controls and PD patients self-reported which emotions they experienced.<br>*Part 2:* observers (age-peers) rated social attributes in video clips (with sound) of PD-patients and controls. | *Part 1:* CPIB: PD: 19.53<br>Raters accuracy was low (10–40%). Women were perceived more accurate. Positive emotions were perceived more accurately in women of control group, compared with women with PD. Negative emotions were more accurately perceived in PD-patients, compared with controls.<br>PD-patients: more often wrongly perceived as experiencing a negative emotion.<br>PD-patients: more facially expressive than controls.<br>*Part 2:* PD-patients were rated as less involved*, interested*, friendly*, intelligent*, optimistic*, attractive*, and attentive*. |
| Tickle-Degnen<br>[38]<br>2004<br>USA | Qualitative observational study<br>PHC and SHC | PD: 6 (83%)<br>Novices: 30 (0%)<br>Experts: 50 (10%) | PD: 49–79 (mdn = 59.5)<br>Novices: 22 (SD = 1.7)<br>Experts: 33 (SD = 7.3) | 2–11 y (mdn = 5.4) | NEO-FFI | Expert (different disciplines) and novice practitioners (students) rated video clips of interviews with PD-patients.<br>Ratings: personality factors.<br>PD patients were rated for facial masking using UPDRS item of facial expressing.<br>Facial masking: high: 2, intermediate: 2, and low: 2. | Correlations with facial masking:<br>Extraversion: novice: r = –0.47*; expert: r = –0.20*.<br>Agreeableness: novice: *r* = –0.15*; expert: *r* = –0.23*<br>Neuroticism: novice: *r* = 0.23*; expert: *r* = –0.22*<br>Openness to experiences: novice: *r* = –0.04; expert: *r* = –0.10<br>Conscientiousness: novice: *r* = –0.10; expert: *r* = –0.05<br>More facial masking: perceived as less extravert and agreeable compared with self-ratings.<br>Neuroticism and conscientiousness: novices were less accurate compared with experts. |
| Tickle-Degnen<br>[39]<br>2011<br>USA | Qualitative observational study<br>SHC and THC | PD: 24 (50%)<br>Practitioners:<br>USA: 156 (12%)<br>Taiwan: 128 (27%) | PD: 67.5 (SD = 8.1)<br>Practitioners:<br>Clinicians (*n* = 159): 31.9 (SD = 8.3)<br>Students (*n* = 125): 22.4 (SD = 3.0) | *m* = 5.7 y (SD = 3.2) | NVQ<br>GDS<br>TIPI | Practitioners rated video-clips (filtered for speech) with PD-patients. Judgements on following variables: depression, sociability, social supportiveness, and cognitive competence.<br>Patients: categorized in low and high facial masking (UPDRS score for facial masking was used due a median split for gender and culture group). | Practitioners: higher masking: more depressed (*r* = 0.96*), less sociable (*r* = 0.89*), less socially supportive (*r* = 0.65*), and less cognitively competent (*r* = 0.77*). This was more prominent in women than men*.<br>Taiwanese practitioners: cognitive competent scores were less favourable in PD patients with higher facial masking compared with American practitioners.<br>American practitioners: sociability scores were less favourable in PD patients with higher facial masking compared with Taiwanese practitioners. |

(*Continued*)

**Table 1.** Continued.

| First author Year Country | Study design Level of health care | Participants | | | Assessment tools psychosocial functioning | Methods | Psychosocial outcomes |
|---|---|---|---|---|---|---|---|
| | | Sample size (% male) | Mean age (years) | Time since diagnosis | | | |
| Wootton [40] 2019 NZL | Qualitative study PHC and SHC | PD: 9 (89%) Partner: 9 (11%) | PD: 40–49: *n* = 1 50–59: *n* = 2 60–69: *n* = 5 70–79: *n* = 1 Partner: 30–39: *n* = 1 50–59: *n* = 3 60–69: *n* = 5 | Expressive impairment duration: *m* = 4.7 y | None | Interview topics: experiences of facial masking, individual and relational impact, and coping with masking. | Facial expression: reduced intensity of showing emotions/reduction in size of facial movements. Discrepancy between verbal and non-verbal communication. Unpleasant social interactions: misinterpretation of emotions due to reduced facial expression. Association of lack of facial expression with negative effect. Compensation methods: use of speech, touch, and gesture. Questioning for confirmation of non-verbal communication. Managing misinterpretation of affect. Facial exercises and posing of non-verbal facial expressions. Giving education about facial masking. |

d: days; EMG: electromyography; FSHD: facioscapulohumeral muscular dystrophy; IHD: ischemic heart disease; *m*: mean; max.: maximum; MD: myotonic dystrophy; mdn: median; mth: months; NVQ: non-validated questionnaire; PD: Parkinson's disease; PHC: primary health care; QoL: quality of life; SD: standard deviation; SHC: secondary health care; THC: tertiary health care; y: years.

Outcome assessment tools used in any of the included studies: AFS: Angstfragebogen für Schüler: 50-item instrument. This is a German anxiety questionnaire for children (age 9–16 years). AT: Abrams and Taylor Scale: assessment tool emotional blunting. Scores range from 0 to 30. BFNE-II: Brief Fear of Negative Evaluation Scale: 12-item instrument, which measures fear and negative evaluation by others. Scores range from 12 to 60 points. Twenty-five point or higher is an indication for social anxiety. COVI: Covi Brief Anxiety Scale: this is an assessment tool for anxiety symptoms. Scores range from 0 to 9. CPIB Short Form: Communicative Participation Item Bank: 10-item instrument, which measures interference of community-dwelling adult with speech related disorders in conversations. Total score ranges from 0 to 30, with higher score as more favourable. DIKJ: Depressionsinventar für Kinder und Jugendliche: 26-item instrument. This is a German version of the Children's Depression Inventory. EHD: Depressive Mood Scale: 20-item scale. Ten items assessing emotional changes expressed by patients and 10 items assessing emotional state by the investigator. FaCE Scale: facial clinimetric evaluation: consists of 15 items with six domains concerning quality of life of patients with facial palsy. Domains: eye comfort, facial comfort, facial movement, lacrimal control, oral function, and social function. Scores range from 0 to 100, with 100 as best score. FDI: Facial Disability Index (FDI): consists of two domains, namely physical function, and social/well-being function. Scores are: 0–100, with 100 as best score. FDQ: Functional Disability Questionnaire: 27-item scale for measuring disability in daily functioning. Scores range from 0 to 108. Higher scores are an indication for more disability in daily functioning. FECQ: Facial Expression Communication Questionnaire: 7-item instrument, which measures the ability to communicate an emotion. A higher score is more favourable. FSI: Facial Image Scale: 13-item instrument, which measures someone's feelings about features and areas of the face. Scores range from 13 to 65. Higher scores are an indication for more positive facial self-assessment. GDS: Geriatric Depression Scale: 15-item instrument. It is a screening assessment for depression in elderly. ≥6 points are an indication for a possible depression. GHQ: General Health Questionnaire: 30-item instrument, which measures anxiety and depression. Scores range from 0 to 30. Higher scores are an indication of more severe anxiety and depression. HADS: Hospital Anxiety and Depression Scale: 14-item instrument, which measures anxiety and depression. Scores range from 0 to 21 points. ≤7 points correlates with no depression or anxiety, 8–10 points with minor depression or anxiety, 11–15 with moderate depression or anxiety, and ≥16 points with severe depression or anxiety. HB-Score: House–Brackmann Score. This is a facial grading system, ranging from I (normal function) to VI (total facial paralysis). HDRS: Hamilton Depressive Rating Scale: 17-item instrument, which is a depression assessment scale. Scores range from 0 to 51. Higher scores are indicating a more severe depression. K10: Kessler 10-item Psychological Distress Scale: consists of 10 questions about the frequency of psychological distress. Every question can be scored from 1 (never) to 5 (all the time), which leads to a minimum score of 10 and maximum score of 50. A higher score is an indication of more psychological distress. KINDL is a 24-item instrument, which measures health-related quality of life in children and adolescents. A higher number is more favourable. Life impact score was calculated by giving a number to each response. Scores range from 0 to 4. 0 equals that a patient experience the issue, but it does not affect patients' life. 4 equals a severely impact in patients' life. MADRS: Montgomery and Asberg Depression Rating Scale: 10-item instrument, which measures severity of depressive episodes. Scores range from 0 to 60. Higher scores are indicating a more severe depression. MDS-UPDRS: Movement Disorder Society's Unified Parkinson's Disease Rating Scale: 65-item instrument, which consists of four parts. Namely: nonmotor experiences, motor experiences, motor examination, and motor complications. MOSSSS: Medical Outcomes Study: Social Support Survey: 3-item instrument. It measures partner enjoyment. Higher scores are more favourable. MWB: Mental Well-Being Scale: this is a measurement for mental well-being. Scores range from 0 to 70. NEO-FFI: NEO-Five Factor Inventory: 60-item instrument assessing five personality factors. Namely: neuroticism, extraversion, openness to experience, agreeableness, and conscientiousness. PAS: Physical Anhedonia Scale: this is a 40-item instrument for assessing anhedonia. PDQ-39: Parkinson's Disease Questionnaire: 39-item instrument, which measures eight dimensions of social life of Parkinson's disease patients. Scores range from 0 to 100. Higher scores are less favourable. PFK 9–14: Persönlichkeitsfragebogen für Kinder und Jugendliche: this is a standardised German Personality Questionnaire for Children (9–14 years). The dimension self-perception was used and consists of 60 items. PNSEA: Positive and Negative Social Exchange Assessment: measures support ratings. SAD-Scale: Social Avoidance and Distress Scale: 28-item instrument, which measures anxiety and avoiding of social interactions. Scores range from 0 to 28 points. Higher scores are an indication for more anxiety and social avoidance. SAQ-A30: Social Anxiety Questionnaire for Adults: 30-item instrument, which measures stress and nervousness in social interactions. Scores range from 30 to 150 points. Higher scores are an indication for more stress and nervousness in social interactions. SAS: Social Anhedonia Scale: this is a 48-item instrument for assessing anhedonia. SDQ-Deu: Strengths and Difficulties Questionnaire (German version): this is a 25-item questionnaire for parents of children in age of 4–16 years old. It consists of five themes: emotional symptoms, conduct problems, hyperactivity, peer problems, and prosocial. A higher number indicates more problems. SF-6D: Short From Six-Dimension: scores estimate the value someone assigns to a health state. Scores range from 0.3 to 1.0. 1.0 is an indication for perfect health. SINHP: Social Isolation domain of the Nottingham Health Profile: 5-item instrument. It measures social isolation and loneliness. Lower scores are more favourable. SSCI: Stigma Scale for Chronic Illness: 24-item instrument. It measures stigma, unfavourable attitude of others, anxiety, shame, and selfdiscrimination. Lower scores are more favourable. Sunnybrook Facial Grading Score measures symmetry of facial movements and resting symmetry. Scores range from 0 (total facial paralysis) to 100 (normal function). SWAL-QOL: Swallow Quality-of-Life Questionnaire 44-item instrument, which contains questions about dysphagia related symptoms and impact of swallowing problems on quality of life. SWLS: Satisfaction With Life Scale: 5-item instrument, which measures life satisfaction and global well-being. Scores range from 5 to 35. Higher scores are more favourable. TYR: Tyrer Anxiety Scale: this is an assessment tool for anxiety symptoms. Scores range from 0 to 60. TIPI: Ten Item Personality Inventory: 10-item instrument, which assesses personality. TSBI: Texas Social Behavior Inventory Short Form B: 16-item instrument, which measures social competence and social self-esteem. Scores range from 0 to 64. Higher scores are more favourable.

*Statistical significance.

**Table 2.** PubMed search strategy (1946 to 9 June 2020).

| | |
|---|---|
| *#1: Psychosocial functioning*<br>"psychology"[Subheading] OR "Psychology"[Mesh] OR "Behavioral Symptoms"[Mesh] OR behavio*[tiab] OR emotional aspect*[Title/Abstract] OR social participation[MeSH Terms] OR social participation[Title/Abstract] OR social distanc*[tiab] OR social behavio*[Title/Abstract] OR social behavior[MeSH Terms] OR "Communication"[Mesh] OR communication[Title/Abstract] OR community participation[Title/Abstract] OR community participation[MeSH Terms] OR quality of life[MeSH Terms] OR quality of life[Title/Abstract] OR QoL[tiab] OR HRQoL[tiab] OR life quality[tiab] OR psychological[Title/Abstract] OR "Deglutition Disorders"[Mesh:NoExp] OR "Deglutition"[Mesh] OR Deglutition[tiab] OR swallowing[Title/Abstract] OR speaking[Title/Abstract] OR talking[Title/Abstract] OR personal satisfaction[MeSH Terms] OR well-being[Title/Abstract] OR satisfaction[Title/Abstract] OR occupation[Title/Abstract] OR "Employment"[Mesh] OR professional occupation[Title/Abstract] OR psychosocial*[Title/Abstract] OR social interaction[title/abstract] OR social competenc*[title/abstract] OR social disabilit*[title/abstract] OR social isolation[title/abstract] OR social stigma[title/abstract] OR social adjustment*[title/abstract] OR social discrimination[title/abstract] OR Social Desirability[title/abstract] OR Social Isolation[title/abstract] OR social skill*[title/abstract] OR social attitude[title/abstract] OR mental stress[title/abstract] OR psychological stress[title/abstract] OR relation*[title/abstract] OR wellbeing[title/abstract] OR employment[title/abstract] OR unemploy*[title/abstract] | 4 664 067 |
| *#2A:* Diseases *1*[a]<br>myotonic dystrophy[MeSH Terms] OR parkinson disease[MeSH Terms] OR myotonic dystroph*[Title/Abstract] OR parkinson*[Title/Abstract] OR steinert disease[Title/Abstract] OR steinert's disease[Title/Abstract] OR steinert myopath*[Title/Abstract] OR steinert's myopath*[Title/Abstract] | 130 945 |
| *#2B: Diseases 2*[a]<br>bell palsy[MeSH Terms] OR mobius syndrome[MeSH Terms] OR bells pals*[Title/Abstract] OR mobius syndrome[Title/Abstract] OR moebius syndrome[Title/Abstract] OR muscular dystrophy facioscapulohumeral[MeSH Terms] OR fshd*[Title/Abstract] OR facioscapulohumeral muscular dystrophy[Title/Abstract] OR bell pals*[Title/Abstract] OR bell's pals*[Title/Abstract] | 4938 |
| *#3: Altered facial expression*<br>facial expression[MeSH Terms] OR facial paralysis[MeSH Terms] OR facial weakness[Title/Abstract] OR facial expression[Title/Abstract] OR facial paralysis[Title/Abstract] OR hypomimia[Title/Abstract] OR facial muscles[MeSH Terms] OR mimetic muscle[Title/Abstract] OR facial paresis[Title/Abstract] OR facial muscl*[Title/abstract] OR facial nerve paralysis[Title/abstract] | 37 868 |
| #1 AND #2A AND #3 | 295 |
| #1 AND #2B | 618 |
| (#1 AND #2A AND #3) OR (#1 AND #2B) | 909 |
| (#1 AND #2A AND #3) OR (#1 AND #2B) + filter: language: English and Dutch | 811 |

[a]We identified two categories of diseases. The first category: diseases with altered facial expression, but not as the main symptom (Parkinson's disease and myotonic dystrophy type 1). The second category: diseases with altered facial expression as one of the major symptoms of disease (FSHD, Bell's palsy, and Moebius syndrome).

difficulties could only be included when also having psychosocial outcome measures.

Quantitative studies could be selected if the study incorporated a tool to measure the outcomes of interest. For qualitative studies together, these aspects provide a broad perspective of psychosocial functioning. Studies were excluded when only the psychosocial consequences of other symptoms than altered facial expression were investigated.

### *Search strategy*

The databases of PubMed (1946 to 9 June 2020), Embase (1974 to 9 June 2020), and the Cochrane Library (1966 to 9 June 2020) were searched. There was no restriction in publication date, since older studies were not expected to be outdated regarding psychosocial outcome measures. An overview of the PubMed search strategy is given in Table 2. Search strategies for the other databases are available upon reasonable request. In addition, references from included articles and reviews were checked for possible inclusion. Articles citing one of the included studies were also assessed for possible inclusion.

### *Study selection*

All articles were screened independently by two assessors (N.R. and W.G.). Initially, a title and abstract screening was undertaken followed by full text screening for the selected articles. Disagreements were solved by discussion between the assessors. If there still was no agreement, the principal investigator (N.V.) was also involved in the discussion until consensus was reached.

### *Quality assessment*

The methodological quality was assessed with the NIH/NHLBI Study Quality Assessment Tool for Observational Cohort and Cross-Sectional Studies [42]. This tool consists of 14 questions, which can be scored with "yes", "no", "cannot determine", "not applicable", or "not reported". Questions answered with yes, scored one point. The maximum score is therefore 14 points. Studies with a higher score correlate with a lower risk of bias. Quality assessment was also conducted independently by two assessors (N.R. and W.G.) and disagreements were solved with discussion. When necessary, the principal investigator (N.V.) was also involved in the discussion until consensus was reached.

### *Data collection*

Data were extracted for study and patient characteristics, and outcome measures. Study characteristics included: authors, year of publication, level of health care, country, and design. Level of health care was extracted because it gives an indication of the study population. Patient characteristics included: number of participants, age (mean and range), diagnosis, and duration of symptoms. Outcome measures included: psychosocial functioning and type of assessment for indicating psychosocial functioning. For both qualitative and quantitative studies, results and general themes were collected and incorporated in Table 1 and the results section. The general themes were discussed in "Discussion" section. A meta-analysis was not performed due to the heterogeneity in diseases and study designs.

### *Data availability statement*

The data that support the findings of this study are available from the corresponding author upon reasonable request.

## Results

### *Search*

The initial literature search resulted in 2298 articles (The Cochrane Library: 137, Embase: 1350, and PubMed: 811). Duplicates were removed and a total of 1629 articles remained. Screening for title and abstract led to 53 remaining articles. The

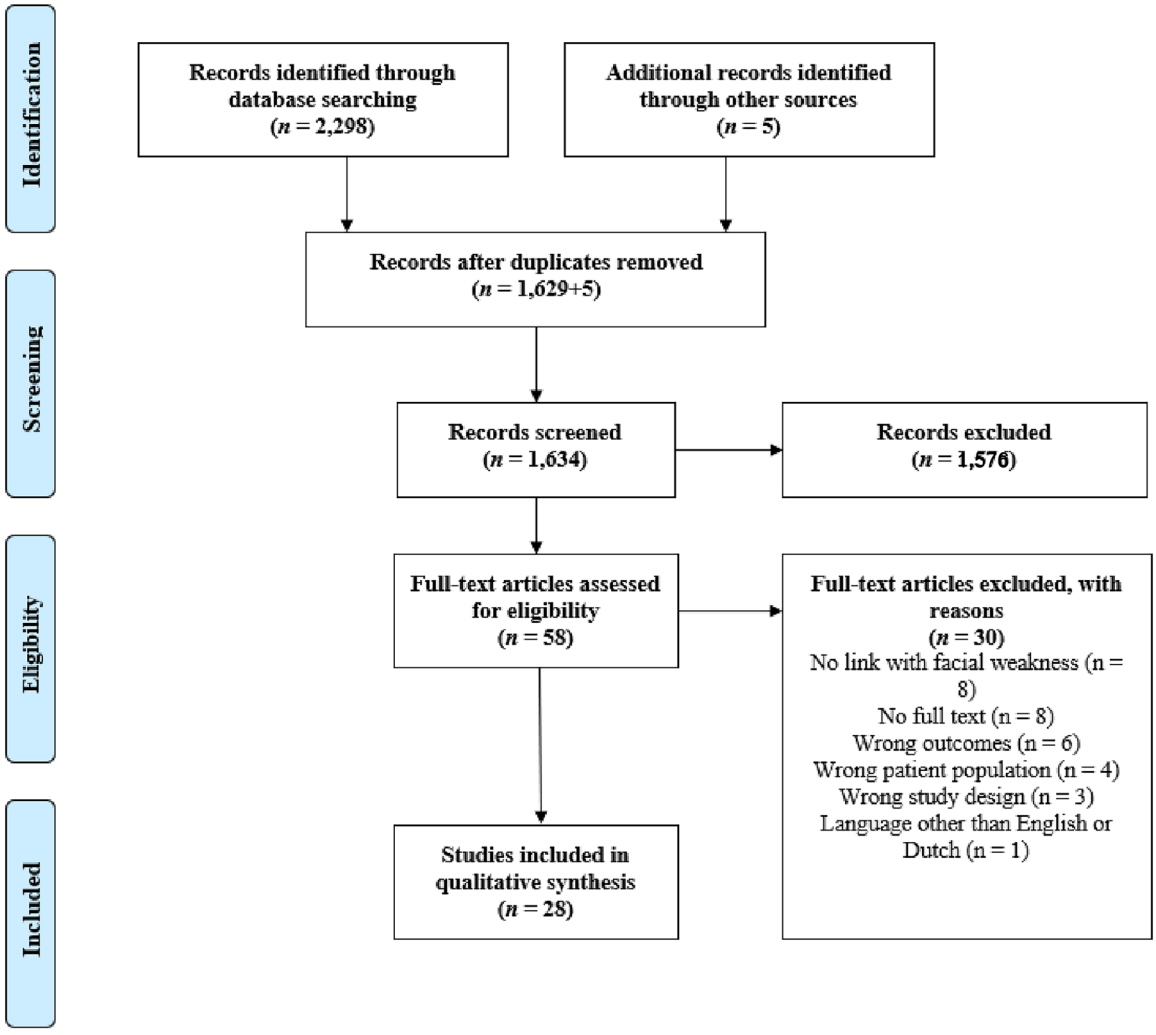


**Figure 3.** PRISMA flowchart for study inclusion.

most frequent reasons for exclusion in title and abstract screening were missing psychosocial outcomes or not containing any of the five neurological diseases of interest. The full-text screening resulted in 25 articles (details in Figure 3). Reference lists of these 25 articles and articles that cited one of the 25 articles were checked for possible inclusion. This led to five additional articles. Three of these articles were eventually included, and two were excluded during full text screening. Thus, in total, 28 articles were identified to meet the inclusion criteria for this review.

### *Study characteristics*

Studies were included on all five diseases of interest. Parkinson's disease and Bell's palsy studies were most frequent, with ten [31–40] and eight [13–20] included articles, respectively. The other diseases were included less: Moebius syndrome with five studies [25–29], FSHD with four studies [21–24], and lastly myotonic dystrophy type 1 with one study [30]. Most studies had a qualitative design (in-depth interviews; $n$ = 4 and observational studies; $n$ = 7) or were quantitative questionnaire studies ($n$ = 6). Studies were performed in four continents (North America, 54%; Europe, 32%; Asia, 11%; Australia, 4%). Sample sizes varied between eight participants [36], and 322 970 participants [17]. The mean age of participants ranged from 11 to 69 years [29, 33]. All studies together used 42 different tools to assess psychosocial functioning, including disease specific tools and tools in a language other than English. Further characteristics of all studies are summarized in Table 1.

### *Quality assessment*

Most studies (79%) scored 7–9 points of the maximum score of 14 points for quality assessment. Main limitations of the included studies were: no reported participation rate of eligible patients, no sample size justification, and no or too little adjustments for potential confounding variables. Only three studies [22, 26, 29] reported the participation rate of eligible patients. The minimal rate of participation was 54% (range 54–100%). Nineteen studies had not performed a sample size justification. There were sufficient adjustments for potential confounding variables in 10 studies. In only one study [13], an exposure assessment was done more than once. Two questions of the used quality assessment tool [42] were not applicable in most studies. These questions assessed blinding of outcome assessors to exposure status, and follow-up rate. These questions were mostly unapplicable because of the used study design. An overview of quality assessments of all studies is shown in Table 3.

### *Study outcomes*

All study outcomes are presented per neurological disease. A total overview of outcome measures, including the study designs, is shown in Table 1. The main findings are summarized below.

**Table 3.** Quality assessments of included studies (NIH/NHLBI study quality assessment tool for observational cohort and cross-sectional studies).

| | 1. Research question | 2. Study population | 3. Participation rate | 4. Eligibility criteria | 5. Sample size | 6. Exposure measures prior to outcome | 7. Time frame | 8. Different level of exposure | 9. Exposure definition | 10. Exposure assessment frequency | 11. Outcome measures | 12. Blinding outcome assessors | 13. Follow-up | 14. Confounding variables | Overall score |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Bakker et al. [21] | Yes | Yes | NR | Yes | Yes | Yes | Yes | Yes | No | No | Yes | NR | NA | Yes | 9 |
| Bogart and Matsumoto [25] | Yes | Yes | NR | Yes | No | Yes | Yes | Yes | No | No | Yes | NA | NA | No | 7 |
| Bogart et al. [26] | Yes | Yes | Yes | Yes | Yes | Yes | Yes | No | Yes | No | Yes | NR | NA | No | 9 |
| Bogart [27] | Yes | Yes | NR | Yes | NA | Yes | Yes | Yes | Yes | No | Yes | No | NA | No | 8 |
| Briegel [28] | Yes | Yes | NR | Yes | No | Yes | Yes | Yes | Yes | No | Yes | NA | NA | No | 8 |
| Bungener et al. [30] | Yes | Yes | NR | Yes | No | Yes | NR | Yes | Yes | No | Yes | Yes | NA | No | 8 |
| Bylund et al. [13] | No | Yes | NR | Yes | No | Yes | Yes | Yes | Yes | Yes | Yes | NA | Yes | Yes | 10 |
| Gunnery et al. [31] | Yes | Yes | NR | Yes | No | Yes | NR | Yes | Yes | No | Yes | NA | NA | Yes | 8 |
| Hamel et al. [22] | Yes | Yes | Yes | Yes | No | Yes | Yes | Yes | Yes | No | Yes | NA | NA | Yes | 10 |
| Hemmesch et al. [32] | Yes | No | NR | Yes | No | Yes | Yes | Yes | Yes | No | Yes | NA | NA | No | 7 |
| Hemmesch [33] | Yes | Yes | NR | Yes | Yes | Yes | Yes | No | Yes | No | Yes | NA | NA | Yes | 9 |
| Huang et al. [14] | Yes | Yes | NR | Yes | NR | Yes | No | Yes | Yes | No | Yes | NA | NA | Yes | 8 |
| Johnson et al. [23] | Yes | NR | NR | Yes | No | Yes | NR | NR | No | No | Yes | NA | NA | NA | 4 |
| Kang et al. [34] | Yes | Yes | NR | Yes | Yes | Yes | Yes | No | Yes | No | Yes | NA | NA | No | 8 |
| Kim et al. [15] | Yes | Yes | NR | Yes | No | Yes | Yes | Yes | No | No | Yes | NA | NA | Yes | 8 |
| Krane et al. [16] | Yes | Yes | NR | Yes | No | Yes | Yes | Yes | No | No | No | NA | NA | Yes | 7 |
| Lee et al. [17] | Yes | Yes | NA | Yes | Yes | Yes | Yes | No | No | No | Yes | NA | NA | Yes | 8 |
| Ma et al. [35] | Yes | Yes | NR | Yes | No | Yes | Yes | Yes | Yes | No | Yes | NA | NA | Yes | 9 |
| Mul et al. [24] | Yes | Yes | NR | No | No | Yes | Yes | Yes | Yes | No | Yes | NA | NA | No | 7 |
| Pentland et al. [36] | Yes | Yes | NR | No | No | Yes | Yes | No | No | No | Yes | NA | NA | No | 5 |
| Pouwels et al. [18] | Yes | Yes | NR | Yes | Yes | Yes | Yes | Yes | Yes | No | Yes | NA | NA | No | 9 |
| Schwartz and Pell [37] | Yes | Yes | NR | Yes | No | Yes | Yes | No | No | No | Yes | NA | NA | Yes | 7 |
| Smith et al. [19] | Yes | Yes | NA | No | No | Yes | Yes | No | No | No | No | NA | NA | No | 4 |
| Strobel and Renner [29] | Yes | Yes | Yes | Yes | Yes | Yes | Yes | No | No | No | Yes | NA | NA | No | 8 |
| Tickle-Degnen and Lyons [38] | Yes | Yes | NR | Yes | No | Yes | Yes | Yes | Yes | No | Yes | NA | NA | No | 8 |
| Tickle-Degnen et al. [39] | Yes | Yes | NR | Yes | No | Yes | Yes | Yes | Yes | No | Yes | NA | NA | No | 8 |
| Weir et al. [20] | Yes | Yes | NR | Yes | No | Yes | Yes | Yes | Yes | No | Yes | NA | NA | Yes | 9 |
| Wootton et al. [40] | Yes | Yes | NR | Yes | No | Yes | Yes | No | No | No | Yes | NA | NA | No | 6 |

NA: not applicable; NR: not reported.

Questions used in the NIH/NHLBI Study Quality Assessment Tool for Observational Cohort and Cross-Sectional Studies.

1. Was the research question or objective in this paper clearly stated?
2. Was the study population clearly specified and defined?
3. Was the participation rate of eligible persons at least 50%?
4. Were all the subjects selected or recruited from the same or similar populations (including the same time period)? Were inclusion and exclusion criteria for being in the study prespecified and applied uniformly to all participants?
5. Was a sample size justification, power description, or variance and effect estimates provided?
6. For the analyses in this paper, were the exposure(s) of interest measured prior to the outcome(s) being measured?
7. Was the timeframe sufficient so that one could reasonably expect to see an association between exposure and outcome if it existed?
8. For exposures that can vary in amount or level, did the study examine different levels of the exposure as related to the outcome (e.g., categories of exposure, or exposure measured as continuous variable)?
9. Were the exposure measures (independent variables) clearly defined, valid, reliable, and implemented consistently across all study participants?
10. Was the exposure(s) assessed more than once over time?
11. Were the outcome measures (dependent variables) clearly defined, valid, reliable, and implemented consistently across all study participants?
12. Were the outcome assessors blinded to the exposure status of participants?
13. Was loss to follow-up after baseline 20% or less?
14. Were key potential confounding variables measured and adjusted statistically for their impact on the relationship between exposure(s) and outcome(s)?

### Bell's palsy

Eight studies with Bell's palsy patients were included, of which four also included other causes of facial palsy. The proportion of Bell's palsy patients in those studies was: 54% [15], 63% [16], 51% [18], and 75% [19]. Two studies assessed the differences in quality of life for Bell's palsy and other causes of facial palsy [15,16]. Both studies used the Facial Clinimetric Evaluation Scale (FaCE Scale), which is a self-assessment tool concerning quality of life of patients with facial palsy. Patients in Krane et al. [16] with iatrogenic, neoplastic, or traumatic cause of facial palsy had lower quality of life, compared with idiopathic or infectious cause (FaCE Scale total score was respectively 44.78, and 66.13, with a higher score indicating better quality of life). In Kim et al. [15], Bell's palsy patients scored better on FaCE subscale facial movement, but worse on eye comfort and facial comfort subscales, compared with neoplastic aetiology.

Three studies compared psychosocial outcomes between patients and healthy controls. Pouwels et al. [18] assessed anxiety and depression with the Hospital Anxiety and Depression Scale (HADS). Facial palsy patients more frequently had scores indicating mild anxiety, and mild and moderate depression. There were no differences for moderate and severe anxiety or severe depression between both groups. In a case control study by Huang et al. [14], a higher psychological distress in patients than healthy controls was found: on the Kessler 10-item Psychological Distress Scale (K10). Furthermore, Bell's palsy patients tended to have an increased risk of developing depression when compared with healthy controls [17]. In a questionnaire study by Smith et al. [19], 70% of patients stated they noticed an effect of facial weakness in social interactions. The researchers did not specify this effect. Changes in peoples' attitudes towards them were noted by 12 of the 20 patients in the study of Weir et al. [20]. These changes were partly negative (unpleasant comments and staring) and partly positive (kind and more sympathetic). This study also looked at functional disabilities with Functional Disability Questionnaire (FDQ), a 27-item questionnaire for assessing disability in daily functioning. Social activities were mostly affected: almost 50% of the study population noticed this as a functional disability.

The correlation between severity of facial palsy and psychosocial functioning was explored in two studies. Bylund et al. [13] found a correlation of 0.27, which they considered a fair to moderate correlation between social functioning (FaCE Score Subscale: Social Function) and objective facial function (Sunnybrook Scores). Huang et al. [14] compared psychosocial distress (K10 Scale) in patients with mild facial weakness (House–Brackmann Scale: 1–3) and severe facial weakness (House–Brackmann Scale: 4–6). A significant higher psychosocial distress score (K10 Scale) was seen in patients with severe facial weakness than patients with mild facial weakness. In contrast, Weir et al. [20] found only a low association between severity of facial palsy and changes in psychosocial functioning.

Differences between men and women were seen in several studies [13,14, 16,17]. FaCE Social Subscale and Facial Disability Index (FDI) social/well-being scores were both lower for women, compared with men with similar Sunnybrook Scores in Bylund et al. [13]. In the study of Huang et al. [14], psychological distress was higher in women than in men. Psychological distress was measured with K10; although significant, this difference was small (28.07 vs. 26.38). Krane et al. [16] found higher scores for women in fear of negative evaluation (Brief Fear of Negative Evaluation, BFNE-II) and lower FaCE total scores in women, whereas they found no differences for health state (Short From Six-Dimension, SF-6D), social anxiety (Social Anxiety Questionnaire for Adults, SAQ-A30 and Social Avoidance and Distress Scale, SAD-Scale) and social avoidance (SAD-Scale). In a South-Korean nationwide cohort study by Lee et al. [17], women ≥40 years had an increased risk in developing depression. There was no increased risk for women in the <40 years category.

### FSHD

Two of the four included studies with FSHD patients performed interviews to identify the impact of FSHD on someone's life. Bakker et al. [21] found that facial weakness can be experienced as a burden, because the visibility in mirrors and pictures always confronts patients with their disease. In addition, it also negatively interferes with non-verbal communication. If patients talked to others about their disease, they noted that this led to more understanding of earlier noticed limitations and to fewer negative emotions, such as shame and guilt. Johnson et al. [23] asked patients to identify the symptoms that had the most impact on their lives. This led to 251 identified symptoms. Social role limitations were mentioned more often than facioscapulohumeral weakness. These limitations were not only caused by facial weakness. Twenty-five percent of the patients mentioned facial weakness, the inability to smile, and impaired facial expression as one of the symptoms with the most impact. In a cross-sectional study by Hamel et al. [22], patients reported the prevalence and importance of symptoms. Communication difficulties were noted by one-third of the study population. In contrast, problems with shoulders or arms were noted by 97% of patients. Most patients noted consequences of the disease in social interactions, namely decreased performance (77%) and satisfaction (72%) in social situations. Hamel et al. also found a difference between men and women in the impact score for decreased social interactions due to FSHD. Men had an impact score of 1.48 and women 1.84, wherein a score of 1 indicates little effect and a score of 2 indicates moderate effect in patients' lives. Mul et al. [24] looked at strength of orofacial muscles, and the frequency of swallowing and communication problems in FSHD patients. The authors found reduced cheek compression strength, swallowing difficulties (Swallow Quality-of-Life Questionnaire, SWAL-QOL) in 25%, and communication difficulties (Communicative Participation Item Bank, CPIB) in 35% of FSHD patients.

### Moebius syndrome

Three of the five included studies on Moebius syndrome were performed in children (aged <18 years) only [27–29]. Two different qualitative studies of Bogart performed interviews with multiple patients, one in children [27] and the other study in adults [26]. These studies point out that some patients compensate the lack of facial expressions with various strategies: tone of voice, body movements, humour, and speaking slowly, loud, and clearly to avoid stuttering. Several topics came up for discussion in both children and adults: lack of public awareness, social disengagement, stigma, and being misunderstood. Patients noticed that others often do not know how to react when they are not aware of Moebius syndrome. Speech, expression of emotions, and showing your personality are all more difficult due to facial weakness. Social strain, positive outlook, and aggression were only mentioned by children, while discrimination was only mentioned by adults.

Briegel [28] and Strobel and Renner [29] both looked at differences between patients and their parents concerning psychosocial functioning. Anxiety (*Angstfragebogen für Schüler*, AFS),

depression (*Depressionsinventar für Kinder und Jugendliche*, DIKJ), and self-perception subscales (*Persönlichkeitsfragebogen für Kinder und Jugendliche*, PFK 9–14) were lower than or similar to normative data [28]. Similar findings were seen in Bogart and Matsumoto [25], as scores for anxiety, depression (HADS), and life satisfaction (Satisfaction With Life Scale, SWLS) were not significantly different from normative data. However, in the same study, they found that social competence (Texas Social Behavior Inventory Short Form B; TSBI), and emotion expression were lower than normative data. Parents rated their children more often as having more social or emotional problems (Strengths and Difficulties Questionnaire, SDQ) compared with self-reports of the children [28,29]. Strobel and Renner [29] found that health related quality of life (Questionnaire for Measuring Health-Related Quality of Life in Children and Adolescents, KINDL) was not reduced, with exception for the subscale "friends".

### *Myotonic dystrophy type 1*

Only one study by Bungener et al. [30] on myotonic dystrophy met the inclusion criteria. This French study compared psychosocial impairment in patients with FSHD, myotonic dystrophy type 1, and healthy controls. Anxiety (Covi Brief Anxiety Scale; COVI, and Tyrer Anxiety Scale; TYR) and social anhedonia (Social Anhedonia Scale; SAS) were not significantly different between the groups. Myotonic dystrophy patients had significantly lower scores in one anxiety assessment tool (TYR). Emotional blunting (reduced affect display and anhedonia) and emotional deficits (lack of emotional initiative, affective monotony, and anhedonia) were most present in myotonic dystrophy patients. Patients who had emotional deficits had no symptoms of depression or anxiety, unlike FSHD patients with emotional deficits who were more depressed and anxious. One myotonic dystrophy patient met criteria for a depressive state, and two FSHD patients met criteria for a dysthymic state. FSHD patients scored highest in two different depression assessment tools, Montgomery and Asberg Depression Rating Scale (MADRS), and Hamilton Depressive Rating Scale (HDRS), although these scores were only significant when compared to healthy controls. Myotonic dystrophy and FSHD patients had both significantly higher scores for Physical Anhedonia Scale (PAS) when compared with healthy controls.

### *Parkinson's disease*

Seven out of ten included studies with Parkinson's disease patients were observational studies with observers rating patients for social skills and emotional states [32–34, 36–39]. Parkinson's disease patients were perceived as less attractive, less friendly, less interested, and less involved than healthy controls in Schwartz and Pell [37]. Furthermore, Parkinson's disease patients were perceived as more facially expressive. Negative emotions were more accurately perceived and positive emotions were less accurate perceived in Parkinson's disease patients compared with healthy controls. Parkinson's disease patients were also more often wrongly perceived as experiencing a more negative emotion than they really experienced [37]. Pentland et al. [36] compared first impressions on Parkinson's disease patients with ischemic heart disease patients. Parkinson's disease patients were perceived as more anxious, angry, bored, sad, suspicious, and tensed. Their personalities were perceived as more introvert, passive, dependent, and dissatisfied. Although these patients scored not aberrant in psychological tests (similar to the control group and in a non-pathological range). A South Korean study by Kang et al. [34] found reduced facial mimicry for emotional expressions in both spontaneous and voluntary situations.

Judgments about patients with more severe facial masking were more negative, when compared with patients with less severe facial masking. Namely, patients with more severe facial masking were rated as more depressed [39], less socially supportive [33, 39], less extravert [38], less agreeable [38], and less desirable to form a relationship with [33]. These judgments were more unfavourable for women, since women with more severe facial masking were rated as less socially positive [32,33]. In two studies of Hemmesch et al. [32] and Hemmesch [33], observers had less desire for forming a relationship with women with facial masking and rated them as less supportive. These findings contrasted with men with facial masking, where no negative ratings were observed.

In a cross-sectional study by Ma et al. [35], worse quality of life in women was observed compared to men with Parkinson's disease. They also found a positive correlation between self-reported severity of facial masking and worse quality of life. Another cross-sectional study by Gunnery et al. [31] showed that patients with less facial expression had experienced more social rejection. Furthermore, they showed that depression was positively correlated with less facial expressivity.

Lastly, Wootton et al. [40] performed a semi-structured interview study examining the consequences of non-verbal communication impairment due to hypomimia. The following themes were mentioned by Parkinson's disease patients and their spouses. Emotion display was reduced and led to a mismatch between verbal and non-verbal communication, which often resulted in misinterpretation of emotions. Less facial expression was mostly associated with negative effect. Patients used compensation strategies: verbalizing intentions or emotions, asking for confirmation of used non-verbal communication, performing facial exercises and giving education to others.

## Discussion

We will summarize the main findings below. This scoping review showed that patients with Bell's palsy, FSHD, myotonic dystrophy type 1, and Parkinson's disease more often experienced some degree of psychosocial distress compared to healthy controls. In contrast, children with Moebius syndrome had lower scores on depression and anxiety than controls, whereas parents reported that their children had more emotional and social problems.

The onset of altered facial expression might influence coping. Three of the included diseases can be congenital: FSHD, Moebius syndrome, and myotonic dystrophy type 1 [43–45]. Only in Moebius syndrome and in the congenital form of myotonic dystrophy type 1, altered facial expression is clearly visible after birth [44,45]. In early onset FSHD, symptoms can be present in the first year of life (25%), but dysmorphic features due to muscle weakness are not present at birth [43]. The only included study on myotonic dystrophy was without patients with a congenital form. Hence, the studies on Moebius syndrome were the only on congenital altered facial expression. Remarkably, no negative psychosocial outcomes were reported in this group. This might suggest that having a congenital altered facial expression contributes to a more sophisticated or effective coping strategy, instead of having to adapt to a new situation in adulthood. A study of Bogart showed similar results: patients with acquired facial paralysis had less favourable psychosocial outcomes, including more anxiety and depression, when compared to patients with congenital facial paralysis [46]. This hypothesis could be an explanation for the difference seen in psychosocial outcomes between Moebius syndrome and the other included diseases.

In contrast, there is also a potential detrimental aspect in having an altered facial expression early in life. Especially since social media plays an important role in the newest generation and taking pictures and selfies are part of this. When altered facial expression is present during childhood, it might cause a different emotional and social development. This could play a role in early onset FSHD, Moebius syndrome, and congenital myotonic dystrophy. To express emotions, preverbal children rely largely on facial expressions [47]. Lack of facial expressions may have an impact on emotion processing. The facial feedback hypothesis emphasizes that facial movements modulate the intensity of experienced emotions [48]. This could mean that patients who have reduced facial expression experience emotions differently and have problems with social interaction. Only a small number of the included studies investigated the emotional consequences of having an altered facial expression. Further research could investigate the differences in emotional development in children with an altered facial expression and children with a full range of facial expressions.

### *Research gap and recommendations*

Appearance judgements of others is known to influence psychosocial and biological stress processes [49] and can therefore impact the psychosocial functioning of an individual. However, observational studies investigating this have only regularly been performed in Parkinson's disease [32–34, 36–39]. These studies showed that patients with more hypomimia were perceived as more negative and in some studies this effect was even more evident in women [33, 38,39]. Observational studies could be performed in other diseases with an altered facial expression to examine the relation between severity of facial weakness or facial palsy and the appraisal by others.

The relation between severity of altered facial expression and psychosocial functioning varied across the different disorders. Three studies in Bell's palsy patients investigated the correlation between psychosocial outcomes and severity of facial palsy. Two studies [13,14] found a correlation, whereas one study [20] did not. This is in line with the outcome of studies with various types of facial palsy (Bell's palsy population less than 50%): two American studies showed a correlation between severity of facial palsy and psychosocial outcomes, namely with a higher chance for positive screening for depression [50] and lower quality of life [51]; whereas this was not detected in two English studies [52,53]. Together these studies show that patients with less severe facial palsy can experience impaired psychosocial functioning and patients with more severe facial palsy do not necessarily experience severe impairments. A recent questionnaire study of van de Geest-Buit et al. [54] showed that FSHD patients with more severe facial weakness or facial dysfunction scored lower on different psychosocial outcomes: i.e., fear of negative evaluation and social functioning. This correlation was seen for the self-reported facial weakness scores and not for the physician reported facial weakness scores. Future research could investigate this difference and could also include naive observers and their judgements of psychosocial functioning in FSHD patients with different levels of facial weakness.

Although not a primary outcome, we encountered differences between men and women in multiple studies with Bell's palsy and Parkinson's disease. Both in Bell's palsy [13,14, 16,17] and Parkinson's disease [35], women had more reduced psychosocial functioning than men. Only one included study with FSHD patients by Hamel et al. [22] noted that women had a decreased performance in social situations, but they did not investigate the relation with facial weakness. Further research could investigate if this gender difference is also seen in other diseases with altered facial expression. This possible gender difference is especially important to take into account when potential therapies for improving psychosocial functioning are implemented. Besides the tendency that women, compared with equally affected men, have worse psychosocial functioning, there is also a possible social–cultural phenomena of (consciously or subconsciously) difference in judgement of men and women. Namely, in studies on Parkinson's disease observers appraised patients and the judgements were more unfavourable for women than for men [32,33, 39]. An observational study design can, besides influence of severity of altered facial expression on psychosocial functioning, also assess differences in appraisal between gender and different ages. A possible study design should consist of naive observers who make judgements about personality, social skills, and facial expressivity based on videotaped social interactions between patients with altered facial expression and controls. This observational study would preferably include patients with variable severity of altered facial expression, various ages, and both men and women, to assess these differences. All 10 included studies with Parkinson's disease patients had a relatively old population (mean range: 53–69 years [33, 36]). Young onset Parkinson's disease [55] is not included in these studies. Research has shown that patients with young onset Parkinson's disease have lower quality of life compared with late onset Parkinson's disease patients [56–58]. These studies did not investigate the influence of hypomimia as a possible confounder. It would be interesting to know if the psychosocial impact of hypomimia is different in young onset Parkinson's disease.

Learning compensatory expressive strategies is a potential intervention method for patients with altered facial expression. In one study [40] with Parkinson's disease patients and two studies [26,27] with Moebius syndrome patients, participants reported that over time they had learned how to use compensation strategies (see also Figure 1). This is consistent with one study that found that patients with congenital facial palsy used more compensatory expressive behaviour (usage of body and vocal expression and emotion words) when compared with patients with acquired facial palsy [59]. Learning of compensation strategies has proven to be a successful intervention in Moebius syndrome [60]. Similarly, learning of compensation strategies could be beneficial for other patients with an altered facial expression. In a recent questionnaire study in FSHD, we showed that younger FSHD patients experience more psychosocial distress due to facial weakness than elderly FSHD patients [61]. Therefore, compensatory strategies could be especially beneficial for younger patients. This could also apply for myotonic dystrophy, Moebius syndrome, Bell's palsy during childhood and in young onset Parkinson's disease. Compensatory strategies could consist of tone of voice, gestures, verbalizing emotions and feelings, exaggerate facial expressions and informing others about their disease and altered facial expression. These strategies should be customized per disease and person. Tone of voice differences can be difficult for Parkinson's disease patients to change and exaggerating facial expressions are more impactful in unilateral facial palsy than in bilateral facial palsy. On the other hand, verbalizing emotions and feelings, and informing others are applicable for all included diseases. Further research should identify the best suiting strategies per disease. Compensatory strategies could be learned during a social skill workshop (live or online) or with help of applications and have potentials for improving psychosocial functioning.

Comparing the results of this review (multiple diseases with altered facial expression) with a prior systematic review on the psychosocial impact of facial palsy patients only [12], showed both similar as additional findings. Similarities are: (1) a possible gender

effect (women with altered facial expression are more likely to develop psychosocial problems, observed in Parkinson's disease, Bell's palsy and possibly in FSHD) and (2) objective severity of altered facial expression is not directly associated with psychosocial functioning. This study adds: (1) awareness of potential detrimental effects of altered facial expression on emotional development; (2) awareness of the impact of possible negative judgements of others in patients with less facial expression (seen in Parkinson's disease with patients with more hypomimia); (3) the call for observational studies to investigate this in diseases other than Parkinson's disease; and (4) learning compensatory strategies as a possible intervention method, especially for younger individuals.

None of the included studies had incorporated the ICF framework [62]. The ICF framework is a concept were functioning as a person is described. Multiple components are part of it: body functions, activities, participation, environmental and personal factors, and health condition (disease). The framework shows all the interactions between the categories. Usage of this framework could be useful to describe the psychosocial impact of having an altered facial expression, as it is something that is directly visible for others. Therefore, this health condition can directly influence behaviour regarding participation and doing activities due to (fear of) reactions of others, or patients own perspective of themselves.

### *Limitations*

One limitation of this study is that the neurological diseases are heterogeneous in many aspects and therefore outcomes cannot be extrapolated directly. In Bell's palsy patients, the altered facial expression is the main symptom whereas in the other included diseases the altered facial expression is only one of the several symptoms. Also, the course of disease is different ranging from congenital, acute or slowly progressive in later age. We have added studies with facial palsy patients with other aetiologies than Bell's palsy, as long as the Bell's palsy group was more than 50% of the total study population. Therefore, there is a chance of selection bias when the other aetiology influences the psychosocial outcomes. Furthermore, neurocognitive impairment in Moebius syndrome, myotonic dystrophy type 1, and Parkinson's disease can negatively influence quality of life [45, 63–65]. In myotonic dystrophy type 1, around half of the patients have cognitive impairment (e.g., processing speed, working memory, or mental retardation) [63, 65]. Parkinson's disease patients have a complex pattern of cognitive impairments, which is variable between patients. These impairments consist of reduced attention, impaired memory, difficulties in processing visual input, and language impairments [64]. In 9–15% of patients with Moebius syndrome, intellectual disability is present [66]. It is therefore possible that an altered facial expression has less impact on psychosocial functioning in Moebius syndrome, for example, due to less awareness of the possible impact of having an altered facial expression on social interactions. This variety makes a direct comparison of outcomes difficult. However, it does provide an overview, from which recommendations and limitations can be identified to help in the understanding of psychosocial consequences of altered facial expression.

In collaboration with an information specialist of the medical library (O.C.), three large databases were selected: PubMed, Embase, and Cochrane Libraries. The assumption was that these three databases would cover all potentially interesting studies. PsychINFO was not used as a database and therefore studies could have been missed.

Besides altered facial expression, other symptoms can influence psychological functioning, for example, Parkinson's disease patients are more prone to develop depression [67]. Not every included study did focus on the correlation between the degree of altered facial expression and psychosocial function.

Different critical appraisal tools were considered. The NIH/NHLBI Study Quality Assessment Tool for Observational Cohort and Cross-Sectional Studies seems most applicable for all included studies. By choosing this quality assessment tool, there is a possibility that certain quality assessment aspects specific for qualitative studies were not assessed. To adjust for questions that were not relevant for qualitative studies, "not applicable" was noted. There are other quality assessment tools that could have been used that were more focused on mixed methods review.

The different studies used a total of 42 different outcome assessment tools. In an ideal situation, there would be more uniformity in usage of outcome assessment tools to make comparisons between studies more convenient. It would be convenient if there are assessment tools that are usable for similar diseases, for example, neuromuscular diseases. Effect evaluations and comparisons between diseases is then easier.

Furthermore, several studies had relatively small sample sizes, especially in studies with Moebius syndrome and myotonic dystrophy type 1. Quality of evidence of these studies is lower than in studies with more patients, because the chance of potential bias is higher in small sample sized studies.

## Conclusions

Negative effects of altered facial expression on psychosocial functioning are abundant in a range of neurological diseases and are even more prevalent in women. This review demonstrated the high need for future research in this area, and highlights that attention of health care professionals, friends, and family to the psychosocial consequences of altered facial expression is critical. The different perspectives have given insights for future studies focussing on psychosocial consequences of altered facial expression and especially on emotional consequences. Future observational studies on the performance of patients in social interactions, in line with studies in Parkinson's disease, may help to advance our understanding of the psychosocial consequences in disease with altered facial expression. Future research is also needed to better understand the impact of impaired facial expression beyond emotion expression by looking at the impact on the communication of meaning. Studies on learning of compensation strategies may be another fruitful way forward, as this could be beneficial for patients with an altered facial expression, particularly younger patients.

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## Author contributions

Nathaniël B. Rasing: conceptualization, data curation, investigation, formal analysis, methodology, project administration, visualization,

and writing – original draft. Willianne van de Geest-Buit: conceptualization, data curation, investigation, formal analysis, and writing – review and editing. On Ying A. Chan: data curation and writing – review and editing. Karlien Mul: conceptualization and writing – review and editing. Anke Lanser: conceptualization and writing – review and editing. Corrie E. Erasmus: conceptualization and writing – review and editing. Jan T. Groothuis: conceptualization and writing – review and editing. Judith Holler: conceptualization and writing – review and editing. Koen J. A. O. Ingels: conceptualization and writing – review and editing. Bart Post: conceptualization and writing – review and editing. Ietske Siemann: conceptualization and writing – review and editing. Nicol C. Voermans: conceptualization, formal analysis, methodology, supervision, and writing – review and editing.

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## Data availability statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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