

Mental health and coping strategies in families of children and young adults with muscular dystrophies

Alessandra Tesei¹ · Maria Nobile¹ · Paola Colombo¹ · Federica Civati² · Sandra Gandossini² · Elisa Mani¹ · Massimo Molteni¹ · Nereo Bresolin³ · Grazia D'Angelo²

Abstract

Background Living with a progressive disease as muscular dystrophy (MD) can be challenging for the patient and the entire family from both emotional and practical point of view. We aimed to extend our previously published data about mental health in patients with MDs, also investigating coping profiles of both themselves and their parents. Furthermore, we wanted to verify whether psychological adaptation of patients can be predicted by coping strategies, taking also into account physical impairment, cognitive level and socioeconomic status.

Methods 112 patients with MDs, aged 2–32 were included. Their emotional and behavioural features were assessed through parent- and self-report Achenbach System for Empirically Based Assessment questionnaires and Strength and Difficulties Questionnaires. Development and Well-Being Assessment or Autism Diagnostic Observation Schedule were administered to confirm suspected diagnoses. Coping profile of both parents and patients was assessed through the self-administered New Italian Version of the Coping Orientation to the Problems Experienced questionnaire and its relationship with emotional/behavioural outcome was examined in linear regression analyses.

Results High prevalence of intellectual disability and autism spectrum disorders was confirmed in Duchenne MD. Despite the high rate of internalizing symptomatology, we did not report higher rate of psychopathological disorders compared to general population. Parents tend to rely more on positive reinterpretation and less on disengagement coping. Avoidance coping, whether used by parents or patients, and ID, predicted increased emotional/behavioural problems.

Conclusions Psychosocial interventions should address problems of anxiety and depression that people with MDs frequently experience, even through fostering parents' and childrens' engagement coping over disengagement coping.

Keywords Muscular dystrophy · Emotional problems · Behavioural problems · Psychopathology · Coping strategies · Mental health

Introduction

Muscular dystrophies (MDs) are a heterogeneous group of genetic disorders that share characteristics such as progressive muscle weakness, wasting and lack of therapy [1–3]. Although curative therapy is not yet available, in the last decades, the improvement of standard care has led to a considerable increase in patients' life expectancy [4, 5].

The most severe form of MD, Duchenne muscular dystrophy (DMD), is caused by X-linked dystrophin gene mutations and aff about 1 in 3000 males. Typically, symptoms of DMD manifest between 2 and 5 years of life, ambulation is lost by 12 years, and death mostly occurs in the second or third decade of life due to cardiac or respiratory insuffi [6]. Becker MD (BMD) is the allelic

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milder variant of the disease and affect about 1 in 20,000 males. In BMD, muscle symptoms usually have onset in the second decade, walking autonomy may be preserved up to the fifth or sixth decade, and life expectancy is not significantly reduced, unless cardiomyopathy occurs [7].

Limb-Girdle MDs (LGMDs) include at least 8 different forms of autosomal dominant (LGMD1) and more than 20 distinct forms of autosomal recessive (LGMD2) muscular dystrophies. The overall frequency varies within different populations and is estimated around 20–40/10,000,000 [8, 9]. The most frequent presentation is proximal weakness that progressively impairs daily activities; respiratory and cardiac involvements are uncommon [3].

While severity, age of onset, rate of progression and consequent complications and prognosis are variable among these types of MDs [10], common features are genetic etiology, muscular involvement, disease progression, lack of resolutive therapy and physical limitations that compromise autonomy in daily life activities [11]. Along with motor impairment, neurodevelopmental and emotional/behavioural concerns are frequently associated with MDs [12–14]. This has been documented by numerous studies, which reported higher rate of internalizing symptoms, autism spectrum disorder (ASD), attention-deficit hyperactivity disorder (ADHD), intellectual disability (ID) and difficulty with social functioning in children and young adults with MDs compared to general population [14–21]. As affected by chronic, progressive conditions, people with MDs need constant assistance in terms of medical, physical and emotional care and they gradually experience increased dependency on others, most often on relatives [11, 22]. The increasing demands of care on multiple fronts and the physical, social and psychological hardships associated with the disease represent a potential stressor that patients and their families need to cope with and adapt to [23–25].

Caregivers of patients with MDs have been reported to experience moderate-to-high levels of stress, distress and frequent feelings of guilt, low self-esteem, sadness and depression related to the patient's condition [22, 26–29]. However, relatives with adequate coping skills, high self-esteem, and a sense of support from social network perceive lower burden and identify more positive aspects of the caregiving experience [22, 27, 30]. An interesting study conducted among relatives of MD patients revealed that most perceive their caregiving experience as having a positive impact on their lives (in terms of personal growth, resilience, altruism, sharing of the experience), all the more recognized among those who reported more practical difficulties [30]. This suggests that caregiver's perception of their experience as positive strengthens involvement and confidence in their resources, preventing exceeding stress threshold, and ultimately helping to cope with their child illness and

producing beneficial effects on patients in terms of quality of care received and quality of life.

Moreover, previously published studies report a good quality of life among patients with DMD, especially in the psychological health domain, despite the physical burden [31–33], supporting the hypothesis that internal resources—of both patients and their relatives—could moderate the relationship between illness and psychological adjustment.

In its transactional model of stress and coping, within an ecological system's theory perspective, Thompson views chronic illness as a potential stressor to which the child and the family must adapt, and the relationship between illness and the psychological adjustment depends on biomedical, developmental and even psychosocial processes [34]. In this perspective, coping strategies, of parent and of individual, are hypothesized to be part of those processes moderating the illness–adjustment relationship, over and above the contributions of the illness and demographic parameters [35].

Lazarus and Folkman [36] described coping as thoughts and behaviours that people use to manage the internal and external demands of situations that are appraised as stressful.

Coping strategies have been classified conceptually into “primary control engagement coping”, “secondary control engagement coping” and “disengagement coping”. Primary control engagement coping is aimed at directly changing the stressor or the emotional reactions to the stressor. An example of primary control coping targeting the stressor is problem solving, that involves efforts to define the problem, generate alternative solutions, select the best solution and implement the solution. Primary control coping directed to emotional reactions consists in strategies such as emotional expression and emotional communication to others that aim to get moral support, sympathy, understanding and emotional relief. Secondary control engagement coping comprises efforts to adapt to a stressor rather than change it, by strategies as acceptance (e.g., deciding to learn to live with it) and positive reinterpretation (e.g., looking for something good in what is happening, trying to grow as a person because of the experience). Disengagement coping, conversely, is aimed at escaping from the stressor or related emotions and includes passive and maladaptive responses such as avoidance (e.g., turning to other substitute activities to think about it less), denying the stressor existence and substance use [37].

Research has shown that the use of engagement coping is related to positive health outcome and increased quality of life [37], whereas avoidant-oriented coping has been proven to be ineffective for managing long-term stress and is associated with higher psychological distress, including anxiety and depression [37–41].

Individuals differ in their propensity to rely on some coping strategies over others in response to a stressful situation [38]. However, the use of a specific coping strategy is

a function of specific situational factors, other than dispositional, as both of them contribute to determine the subjective cognitive appraisal of threats and resources to cope with the stressor, from which derives coping behaviour. One aspect particularly relevant involved in the process of appraisal of the situation is the perceived controllability of the stressful event. Perceived control refers to the extent to which a person believes that the outcome of an event can be attributed to internal (personal) sources, external (situational/environmental) sources, or to the cause or predictability of an event [42].

Studies have shown that specific types of coping strategies are more or less effective depending upon their fitting with the appraised controllability of the situation. Primary coping is more adaptive in situations perceived highly controllable, whereas secondary coping is beneficial in situations as chronic illnesses, which, even though perceived as relatively unchangeable and uncontrollable, can be appraised as a call to adapt or as a way to rediscover themselves and the others, leading to a reduction of perceived stress. The flexibility of appraisals of controllability and the flexibility of coping consequent to the appraisal could help the individual's well being and adaptation to the stressful situation, even when facing a severe chronic disease such as MD.

To our knowledge, there is only one past study conducted among patients with MDs examining the relationship between the use of certain coping strategies by parents or patients and behavioural outcomes of these latter. Thompson and colleagues [43], involving children with DMD and their relatives, found that a high use of disengagement over engagement coping by parents was associated with higher levels of both internalizing and externalizing behavioural problems in children. Consistently, more recent studies conducted on other chronic conditions (e.g., diabetes, chronic pain, cancer) have shown substantial evidence that disengagement coping is related to poorer adjustment [40].

In the present study, we aim to (1) extend our previous published data on prevalence of emotional, behavioural and neurodevelopmental problems of children with DMD [15], through the inclusion of other forms of MDs, namely BMD and LGMD together with DMD, and through the extension of age range (we recruited children and young adults until 32 years of age). Because of the progressive nature of the diseases, extending the age range can allow to observe the characteristics of emotional-behavioural profile in relation to increased levels of physical impairment and clinical severity and complexity. The inclusion of different forms of MDs, on the other hand, allows to observe the prevalence of emotional and neurodevelopmental disorders across MDs that have different levels of central nervous system (CNS) involvement (more severe for DMD, milder for BMD, typically absent for LGMD); (2) verify whether there are significant differences in the coping styles of parents and patients

when compared to published general population norms. To our knowledge, the only Italian tool on coping having published norms able to be compared was validated in an adult sample; this constrained our investigation of self-report coping styles, because, to make balanced comparisons, we needed to exclude children. We, therefore, involved all subjects older than 16, as at this age, logical reasoning, planning and other high-order thinking are considered at adult level of maturity [44]; (3) examine the role of parental coping strategies in relation to emotional/behavioural problems of their children; (4) examine the role of patients' coping strategies in relation to their self-reported emotional/behavioural problems. In achieving the latter two aims, the contribution of patients' physical condition and cognitive level will be also taken into account.

It was hypothesized that patients with MDs would display—compared to general population—higher prevalence of neurodevelopmental disorders (i.e., ID, ASD, ADHD), especially for those MDs with a higher involvement of CNS (DMD and BMD). Based on our previous study [15], it was also hypothesized the presence of a higher rate of reported emotional/behavioural symptoms that, however, would not reach diagnosis level. Looking at coping styles, it was hypothesized that both patients and parents would have—compared to general population—a coping profile that would involve a greater use of acceptance and a lower use of problem solving, as typically happen when people adapt to relatively uncontrollable stressors. Finally, we hypothesized that increased physical/cognitive impairment and the use of avoidance strategies would be associated with increased emotional/behavioural problems in patients.

Materials and methods

Participants

A clinical population of 609 patients with inherited neuromuscular disorders, comprising, among these, 133 with DMD (age range 2.8–32), 56 with BMD (age range 7–63) and 70 with LGMD (age range 9–73) attends the Neuromuscular Unit of Scientific Institute IRCCS E.Medea for periodic clinical assessment. Among them, a total of 115 responding to the inclusion criteria (described below) were invited to take part to the study during their attendance at the Institute as inpatients or outpatients. A total of 113 including families or individual patients in case of adults living without parents agreed to participate in the research. The final sample consisted of 112 patients who returned questionnaires. Inclusion criteria were (1) confirmed clinical and molecular diagnosis of DMD, BMD and LGMD, according to international criteria [45–47], (2) age \leq 32 years old, (3)

comprehension of the procedures and aims of the study, (4) signature of a written informed consent form (obtained from parents for patients younger than 18 years).

The study was approved by the Ethical Committee of the ‘E. Medea’ Scientific Institute according to the Declaration of Helsinki.

Procedure

To assess cognitive, emotional and behavioural features, intelligent test was administered to patients and—if aged at least 11—self-report screening questionnaires about their emotional/behavioural problems were proposed. Parents completed the parallel version of the questionnaires, answering questions about their child emotional/behavioural problems. Patients exceeding clinical cut-offs on screening questionnaires were subsequently evaluated with a clinical structured interview, using its parent, adolescent or adult version. Preschool patients with elevated autism spectrum problems based on questionnaires and patients with a clinical diagnosis of ASD based on diagnostic and statistical

manual of mental disorders (DSM) criteria were further assessed with structured observation based on the Autism Diagnostic Observation Schedule (ADOS).

Coping strategies were examined with specific self-report questionnaires completed by both parents and children (if older than 16 years of age), regarding their own dispositional coping styles.

Name of each questionnaire and diagnostic tool, contents and tested sample are illustrated in Fig. 1.

Measures administered to patients

Demographic and clinical characteristics

Demographic features including participants’ age, family structure, and family socioeconomic status (SES) measured using Hollingshead nine-point occupation scale [48] were collected. Clinically relevant information such as walking ability (classifying patients as ambulant or wheelchair

Fig. 1 Name, content and tested sample of questionnaires and diagnostic tools. *YSR 11–18* Youth Self-Report (11–18 years), *CBCL 1.5–5* Child Behaviour Checklist (1.5–5 years), *CBCL 6–18* Child Behaviour Checklist 6–18 years, *ASR 18–59* Adult Self-Report 18–59 years, *ABCL 18–59* Adult Behaviour Checklist 18–59 years, *SDQ 11–18+* Strength and Difficulties Questionnaire 11–18 years or more, *SDQ 2–18+* Strength and Difficulties Questionnaire 2–18 years or more, *DAWBA* Development and Well-Being Assessment, *COPE-NVI* Coping Orientation to the Problems Experienced—New Italian version, *ADOS* Autism Diagnostic Observation Schedule

Informant-based tools			
Patients psychological status			
	Informant	Tested sample	Contents
YSR 11-18	Self	31 children with MDs	Internalizing (anxiety, depression, somatic complaints) and externalizing (aggressive/rule breaking behaviour) symptoms, attention problems, thought problems, social problems
CBCL 1.5-5	Parent	6 children with MDs	
CBCL 6-18	Parent	55 children with MDs	
ASR 18-59	Self	50 young adults with MDs	Internalizing (anxiety, depression, somatic complaints) and externalizing (aggressive/rule breaking behaviour) symptoms, attention problems, thought problems, intrusive behaviour
ABCL 18-59	Parent	29 young adults with MDs	
SDQ 11-18+	Self	78 children and young adults with MDs	Emotional problems, conduct problems, hyperactivity-inattention, peer problems, prosocial behaviours, overall distress and impairment
SDQ 2-18+	Parent	57 children and young adults with MDs	
DAWBA	Parent/Self	31 children and young adults with MDs	Questions exploring psychiatric diagnoses based on the DSM-V and ICD 10 criteria
Coping strategies			
COPE-NVI	Self	51 children and young adults with MDs; 79 parents	Use of social support, avoidance strategies, positive attitude, problem solving, turning to religion when exposed to stress
Observation-based tools			
Diagnosis of Autism Spectrum Disorders (ASD)			
ADOS		7 children with MDs	Assesses communication, reciprocal social interaction, imagination/creativity, stereotyped behaviours and restricted interests to inform the diagnosis of ASD

bound), ongoing therapies and the presence of heart dysfunction were collected for each group of patients.

Cognitive assessment

Cognitive ability was assessed using the Wechsler Intelligence scales or Griffiths scales as required based on the patient's age.

Psychological status: screening questionnaires

Emotional and behavioural problems with their impact on patients' everyday life were assessed by Achenbach System for Empirically Based Assessment (ASEBA) [49, 50] questionnaires and Strength and Difficulties Questionnaire (SDQ) [51]. The ASEBA comprises a family of instruments widely adopted in research and clinical practice measuring emotional/behavioral problems, which are sorted in internalizing, externalizing and total problems broadband scales, and narrowband scales (cross-informant syndrome scales). Children from 11 to 18 years can complete themselves the Youth Self-Report (YSR) form. Adults from age 18 to 59 can be assessed with the self-report questionnaire Adult Self-Report (ASR). According to ASEBA multicultural manual, *t* scores greater than 69 for syndrome scales and greater than 63 for broadband scales (internalizing, externalizing and total problems) were considered to be in the clinical range. Age-appropriate ASEBA self-report form was given to patients. In our sample, the majority of YSR/ASR scales showed Cronbach's α coefficients ranging from 0.88 to 0.61, except four YSR scales (Somatic Complaints, Social Problems, Thought problems, Rule-Breaking behaviour) and two ASR scales (thought problems, intrusive behaviour) with an α coefficient < 0.60 .

SDQ is a briefer comprehensive questionnaire, which has been used as a screening tool for child psychopathology in both community and clinical settings [52]. It consists of 25 items, covering emotional problems, conduct problems, hyperactivity-inattention, peer problems and prosocial behaviours. The SDQ also include an impact supplement to assess chronicity, overall distress, social and educational impairment and burden on others of the reported problems. The impact score is made up by one item asking about perceived level of distress, and four items asking about the interference that the person's emotional/behavioral problems have with family life, friendships, work or study and leisure activities. SDQ exists in several versions, for different informants (parent, teacher, self) and age ranges (4–17 years old, 11–17 years old, 18 years or more). For each scale, score can be categorized as 'abnormal' if it exceeds a specific clinical cut-point [53]. Similar to ASEBA

questionnaires, the self-report version of SDQ was given for patients aged 11 and over. In our sample, Cronbach's α coefficient of self-report SDQ scales ranged from 0.78 to 0.62, except for Conduct problems and Peer problems scales ($\alpha < 0.60$).

Assessment of prevalence of psychiatric disorders based on a structured clinical interview or protocol observation

The clinical assessment was conducted using the Development and Well-Being Assessment diagnostic interview (DAWBA) [54] or the Autism Diagnostic Observation Schedule (ADOS) [55].

The DAWBA combines a structured part and a semistructured part, and it is designed to generate present-state psychiatric diagnoses based on the DSM-V and ICD 10 criteria for children, adolescents and adults. The structured sections explore the following psychopathological areas: separation anxiety, simple phobia, social phobia, panic disorder with/without agoraphobia, post-traumatic stress disorder, obsessive-compulsive disorder, generalized anxiety disorder, major depression, ADHD, behavioural disorder, and less common disorders. The semistructured part of the interview elicits a verbatim account of any reported problems. The DAWBA has satisfactory validity and inter-rater reliability [54, 56]. The answers to the structured questions from the interviews and questionnaires were fed into a computerized diagnostic algorithm. This algorithm provided six levels of prediction of the probability of a disorder, ranging from very unlikely to probable, the "DAWBA Bands." The subject is assigned to one of six probability bands corresponding to the approximate prevalence in an epidemiological sample [57] ranging from less than 0.1% likely to more than 70% likely (the other thresholds are 0.5%, 3%, 15%, and 50%). Definite diagnosis was assigned relying on both provisional diagnoses generated by computer and transcripts of answers to open-ended questions. The interviews were administered by a trained interviewer to patients aged 11 and over exceeding clinical cut-off on screening questionnaires.

The ADOS is a standardized, semistructured observational assessment used to assess communication, reciprocal social interaction, imagination/creativity, and stereotyped behaviours and restricted interests to inform the diagnosis of ASD. The ADOS is organized into four modules based on the individual's chronological age and expressive language level, ranging from preverbal to verbally fluent. All ADOS assessments were administered and scored by a licensed clinical child psychiatrist.

Coping strategies

Coping strategies were evaluated with the New Italian Version of the Coping Orientation to the Problems Experienced (COPE-NVI), a self-report measure composed of 60 items [58, 59] in which respondents were asked to indicate, on a four-point scale, how often they used a specific coping strategy when they experience substantial stress. Response choices are the following: “I usually do not do this at all”, “I usually do this a little bit”, “I usually do this a medium amount”, and “I usually do this a lot”. Results of confirmatory factor analysis (CFA) indicated five basic dimensions of the instrument: (1) social support, refers to the seeking of instrumental help or emotional comfort, (2) avoidance strategies, includes avoidant coping responses such as denial and behavioural, mental or alcohol–drug disengagement, (3) positive attitude, measures acceptance and positive reinterpretation of the event, (4) problem solving, refers to the use of active, problem-focused strategies such as planning and suppression of competing activities, (5) turning to religion refers to relying on religion and lack of humour. We asked patients if older than 16 years of age to complete the questionnaire. In this subsample, Cronbach’s α coefficients ranged from 0.88 to 0.75 across the five COPE scales.

Measures administered to parents

Psychological status of children: screening questionnaires

ASEBA questionnaires Child Behaviour Checklist (CBCL) for ages 1.5–5 (CBCL 1.5–5) and for ages 6–18 (CBCL 6–18) are designed to be completed by parents of preschool or school-age children, respectively; ASEBA Adult Behaviour Checklist for ages 18–59 (ABCL 18–59) can be completed by parents of children aged 18 and over. SDQ also has its parent-report versions, that can be completed by parents of preschool (2–4 years old), school-age (4–17 years old) or adult (aged 18 and over) children. These tools allow exploring children emotional/behavioural problems from parents’ point of view and are structured as their self-report counterparts described above in terms of scales and cut-off scores.

Age-appropriate ASEBA and SDQ parent-report version was given to parents. In our sample, the majority of CBCL/ABCL scales showed Cronbach’s α coefficient ranging from 0.88 to 0.65, except three CBCL scales (somatic complaints, thought problems, rule-breaking behaviour) and four ABCL scales (thought problems, rule-breaking behaviour, intrusive behaviour and total problems) with an

α coefficient < 0.60 . Cronbach’s α coefficient of parent-report SDQ scales ranged from 0.75 to 0.62, except for conduct problems, peer problems and prosocial behaviour scales ($\alpha < 0.60$).

Assessment of prevalence of psychiatric disorders of children based on a structured clinical interview or protocol observation

The parent-report version of DAWBA interviews was administered by a trained interviewer to parents of patients exceeding clinical cut-offs on screening questionnaires.

Coping strategies of parents

Parental coping strategies were also evaluated with the aforementioned COPE-NVI. We asked parents to complete the questionnaire. In this subsample, Cronbach’s α coefficients ranged from 0.87 to 0.72 across the five COPE scales.

Data analysis

Statistical analysis plan is reported in Fig. 2.

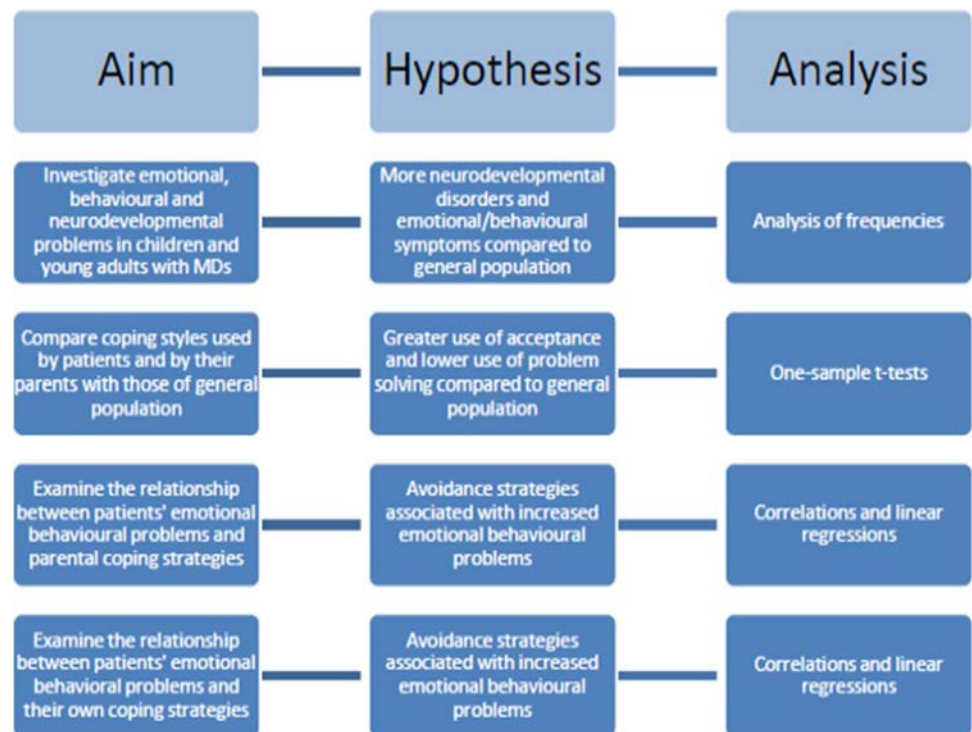
Descriptive statistics were generated for demographic and clinical variables and are reported as mean and standard deviations (SDs) values for continuous variables and frequencies/rates for categorical variables.

For the analysis of emotional/behavioural profile we classified patients in two groups: children (those up to and including 18 years) with their corresponding parent-report measures (CBCL, parent-report SDQ) and adults (those over 18 years), with their corresponding self-report measures (ASR, self-report SDQ). Means and SDs are reported for each scale. The association between SDQ and ASEBA questionnaires was assessed by the Pearson correlation coefficient. Correlations between ASEBA internalizing scale score and the internalizing SDQ scales scores (emotional problems and peer problems) were moderate to strong (ranging from $r = 0.36$ to 0.71). A similar pattern was observed for correlations between ASEBA externalizing scale and SDQ conduct problems scale, which ranged from $r = 0.58$ to 0.61 . ASEBA attention problems scale showed strong correlations with SDQ hyperactivity scale (ranging from $r = 0.73$ to 0.74).

One-sample t tests were performed to compare mean COPE scores of our samples with published Italian norm means [59].

Linear regression analyses were performed to assess the association between coping strategies (COPE scores) and psychopathological symptoms from two perspectives: parental coping strategies in relation to children emotional/behavioural problems derived from parent-report measures CBCL

Fig. 2 Statistical analysis plan



or ABCL (age range 2–31) and patients’ coping strategies in relation to their own self-reported emotional/behavioural problems derived from YSR and ASR (age range 17–32). ASEBA Syndrome Scales *T* scores were used as outcome measures, whereas COPE scales scores were entered as independent variables. In each regression, we included among independent variables only those COPE scales that showed a significant ($p < 0.05$) or a near-significant ($p \leq 0.07$) correlation with a given syndrome scale at the Pearson correlation test (see Supplementary Table 1 and Supplementary Table 2). Similarly, physical impairments, i.e., wheelchair use and cardiac function, were entered as additional independent variables on the basis of previous correlation analysis results. Individual variables age and ID were included by default in the analyses as independent variables. We chose to include in the regression analyses ASEBA rather than SDQ on the basis of the good comparability between measures and of the larger spectrum of emotional/behavioural problems that can be identified by ASEBA.

Bonferroni’s correction was used to adjust for multiple comparisons.

Results

Descriptive results, analysis performed on patients’ sample

Demographic/clinical characteristics and cognitive assessment

Distribution of diagnoses, clinical, demographic features (gender, age, family status, SES) and cognitive level among the 112 patients completing the study are reported in Table 1. Most patients had DMD (75%), followed by BMD (15.2%) and LGMD (9.8%). The mean age of participants was 17.24 (SD 7.46, range 2–32 years). Families of enrolled patients were mainly biparental (82.1%) and they had a medium SES.

At the time of assessment, 72 patients (74.3%) were wheelchair bound, 57 (50.9%) had a cardiomyopathy and, considering the 84 DMD patients, 31 were under current steroid therapy.

The mean full-scale intelligence quotient (FSIQ) was 86.47, with an estimated 22.3% ($n = 25$) of ID (i.e., IQ score below 70); all boys with ID were affected by DMD and they represented 27.9% of the subsample younger than 19 years and 15.7% of the subsample older than 18 years.

Table 1 Clinical and demographic features of the sample ($N=112$)

	Mean (SD)
Age	
Total	17.24 (7.46)
DMD	16.01 (7.34)
BMD	20.24 (7.61)
LGMD	22 (5.04)
VIQ	87.68 (16.90)
PIQ	91.21 (17.64)
FSIQ	86.47 (19.68)
	N (%)
Age range	
Children (2–18 years)	61 (54.50)
Adults (over 18 years)	51 (45.50)
Phenotype	
DMD	84 (75.00)
BMD	17 (15.20)
LGMD	11 (9.80)
Gender	
Males	108 (96.40)
Females	4 (3.60)
Intellectual disability	25 (22.3)
Wheelchair	72 (74.30)
Cardiomyopathy	57 (50.90)
Steroid (only among 84 DMD)	31 (36.90)
	Mean (SD)
Socioeconomic status (SES)	48.29 (19.89)
	N (%)
Family structure	
Two parents	92 (82.10)
Single parent	13 (11.60)
Reconstituted family	3 (2.70)
Live alone	4 (3.60)

SES socio economic status, VIQ verbal intelligence quotient, PIQ performance intelligence quotient, FSIQ full-scale intelligence quotient, DMD Duchenne muscular dystrophy, BMD Becker muscular dystrophy, LGMD limb girdle muscular dystrophy

ASEBA and SDQ

For one adult subject (age 19+), we did not have the self-report scores, as he returned only the parent-report form.

In Table 2, we report mean and SD of CBCL ($N=61$, age 2–18) and ASR ($N=50$, age 19+) syndrome and broadband scales t scores. In Table 3 are reported mean and SD of SDQ parent- ($N=57$, age 2–18) and self-report ($N=50$, age 19+) scale scores. To explore which behavioural outcomes were “clinically relevant”, prevalence and rate of clinical cases for each scale were also reported. Among children,

highest CBCL means emerged for withdrawn/depressed and social problems scales. Looking at broadband scales, a remarkable 24.59% scored in the clinical range for internalizing problems, which is a high rate compared to Italian normative data (9.8% of CBCL caseness according to Frigerio and colleagues [60]). On the contrary, we found very low prevalence of externalizing problems at clinical range (1.64%). Parent completed SDQ mean scores and frequencies of emotional behavioural problems were in line with published data regarding European sample of similar age range [61–63] with the exception of Peer problems scale (17.54% classified as clinical), which showed higher rate of reported symptomatology.

In the young-adult subsample, ASRs profi resembles that of CBCLs as the highest mean score was showed by an internalizing subscale, i.e., Anxious/depressed scale. A high frequency of internalizing problems at clinical range was replicated (18%) and no cases were classified as clinical for externalizing problems, in accordance with prevalence rate of self-reported SDQ clinical conduct problems (0%). A slight elevation of mean score was observed for ASR Attention problems scale, where 2% of young adults fell in the clinical range; this same percentage of clinical cases was also observed for SDQ Hyperactivity scale.

DAWBA and ADOS

Assessment with the clinical structured interview DAWBA was performed on 31 patients out of the total number of 112 (27.68%; 13.4% children, 14.4% young adults) who exceeded clinical cut-off on screening questionnaires. The prevalence of patients actually diagnosed with an emotional and behavioural disorder according to DAWBA was 12.50% (11.48% among children, 13.72% among young adults). In the subsample of children, externalizing disorders were slightly more frequent than internalizing disorders (4.92% of ADHD and 1.64% of oppositional disorders versus 4.92% of anxiety disorders).

Based on protocol criteria, ADOS assessment was conducted with seven children. All of them were DMD and met criteria for ASD, representing 11.48% of the children sample (13.46% of DMD children).

In the young-adult subsample, internalizing disorders were six times more prevalent than externalizing disorders (11.76% of anxiety and depressive disorders versus 1.96% of ADHD); no cases of conduct/oppositional disorder or ASD were found.

Considering DMD, the overall rate of psychopathology was 9.64%. Within the adult DMD subsample, the frequency of psychopathology was 12.5% whereas among DMD children was 7.7%. Regarding the other type of MDs examined, the overall rate of psychopathology was 23.53% within BMD and 18.18% within LGMD.

Table 2 *T* scores (expressed as mean and SD) and prevalence (i.e., number of cases exceeding clinical cut-off) of emotional behavioural problems from ASEBA questionnaires syndrome and broadband (internalizing, externalizing and total problems) scales

	Parent-report CBCL (age 2–18) (<i>N</i> =61)		Self-report ASR (age 19+) (<i>N</i> =50)	
	Mean (SD)	<i>N</i> (%)	Mean (SD)	<i>N</i> (%)
Anxious/depressed	55.98 (6.88)	3 (4.92)	58.12 (7.26)	3 (6)
Withdrawn/depressed	58.38 (8.81)	4 (6.56)	55.12 (5.86)	0 (0)
Somatic complaints	55.95 (5.47)	1 (1.64)	55.64 (6.25)	3 (6)
Social problems	56.28 (6.10)	2 (3.28)	–	–
Thought problems	52.61 (4.24)	1 (1.64)	52.72 (4.29)	0 (0)
Attention problems	53.93 (5.03)	0 (0)	56.48 (5.39)	1 (2)
Rule-breaking behaviour	52.15 (2.84)	0 (0)	52.36 (3.30)	0 (0)
Aggressive behaviour	53.22 (4.52)	1 (1.64)	54.20 (4.45)	0 (0)
Intrusive	–	–	52.12 (3.33)	0 (0)
Internalizing problems	54.50 (10.44)	15 (24.59)	55.10 (10.20)	9 (18)
Externalizing problems	48.30 (8.54)	1 (1.64)	49.32 (7.52)	0 (0)
Total problems	50.35 (10.13)	4 (6.56)	49.90 (8.97)	0 (0)

According to ASEBA multicultural manual, clinical scores correspond to $T > 69$ for syndrome scales and $T > 63$ for broadband scales
CBCL child behaviour checklist, *ASR* adult self-report

Table 3 Raw scores and prevalence (i.e., number of cases exceeding clinical cut-off) of emotional behavioural problems from SDQ questionnaires

	Parent completed SDQ (age 2–18) (<i>N</i> =57)		Self completed SDQ (age 19+) (<i>N</i> =50)	
	Mean (SD)	<i>N</i> (%)	Mean (SD)	<i>N</i> (%)
Emotional problems	1.84 (1.99)	6 (10.53)	2.66 (2.44)	4 (8)
Conduct problems	1.57 (1.50)	6 (10.53)	1.49 (1.23)	0 (0)
Hyperactivity	2.37 (2.25)	3 (5.26)	2.28 (1.94)	1 (2)
Peer problems	1.84 (1.85)	10 (17.54)	1.36 (1.48)	0 (0)
Prosocial	7.67 (1.56)	1 (1.75)	7.87 (1.91)	3 (6)
Total difficulties	7.43 (5.22)	2 (3.51)	7.79 (5.39)	1 (2)
Impact	0.35 (0.85)	7 (12.3)	0.45 (1.16)	6 (12)

Clinical range scores are as follows: emotional problems: 5–10; conduct problems: 4–10; hyperactivity: 7–10; peer problems: 4–10; prosocial: 0–4; total difficulties: 17–40; impact: 2–10

SDQ Strength and Difficulties Questionnaire

Coping strategies

Descriptive analyses for cope scales and statistical comparisons between patients ($N=51$) and previously published Italian data from Sica and colleagues [59] on general community sample are presented in Table 4.

Patients differed from general community as they used less problem solving strategies and lower turning to religion; these differences were still significant after Bonferroni correction; for the other dimensions of COPE patients did not significantly differ from general population.

Descriptive results, analysis performed on parents' sample

Coping strategies

Descriptive analyses for cope scales and statistical comparisons between caregivers ($N=79$) and previously published Italian data from Sica and colleagues [59] on general community sample are presented in Table 4.

Compared to general community, caregivers of patients with MDs relied less on social support, avoidance and problem solving while they had a higher level of positive attitude. After Bonferroni correction, we observed still significant differences for avoidance and problem solving scales.

Correlational analyses

Supplementary Table 1 shows results of correlations between ASEBA parent-report syndrome scales (CBCL/ABCL) and age, SES, physical/cognitive impairment and parents' COPE scales ($N=76$). Supplementary Table 2 shows results of correlations between ASEBA self-report syndrome scales (ASR/YSR) and age, SES, physical/cognitive impairment and patients' COPE scales ($N=51$). Based on these results, regression analysis concerned only those syndrome scales showing a significant ($p < 0.05$) or a near-significant ($p \leq 0.07$) correlation with any COPE scale.

Multivariate analyses

Regressions performed for patient coping

Significant results from multiple linear regression analysis examining the contribution of patients' own coping strategies on their emotional behavioural problems ($N=51$) are presented in Table 5.

The first model statistically significantly predicted anxious/depressive symptoms, explaining 28.1% of the variance. The third model significantly predicted internalizing

Table 4 Mean and standard deviation for the five dimensions of COPE NVI in our groups of caregivers ($N=79$) and patients ($N=51$) compared to general community sample, composed by 457 individuals

	General population sample $N=457^a$	COPE NVI patient $N=51$	COPE NVI parent $N=79$
COPE NVI			
Social support	27.7 (8.4)	26.80 (6.86)	26.03 (7.22)*
Avoidance strategies	23.5 (5.1)	23.57 (5.05)	21.73 (4.90)**
Positive attitude	30.9 (6)	31.43 (6.27)	32.38 (5.96)*
Problem solving	32 (6.7)	27.51 (6.42)**	30.10 (6.25)**
Turning to religion	22.7 (5.6)	19.86 (4.70)**	22.99 (4.67)

COPE NVI Coping Orientation to the Problems Experienced—New Italian version

* $p \leq 0.05$ compared to mean of general community sample

**Significant difference after Bonferroni correction ($p \leq 0.01$; $0.05/5$)

^aSica et al. [59]

problems, and overall accounted for 30.0% of the variance. In both models, avoidance coping strategies were significant predictors of the outcome (and remained marginally significant after Bonferroni correction), with a positive Beta coefficient. Other predictors were not significantly related to the outcome.

Aggressive behaviour as dependent variable was examined in the second model, which explained 24.6% of the variance in this scale. Avoidance coping strategies were

significant predictors of aggressive behaviour scores (even after Bonferroni correction), with a positive Beta coefficient.

The fourth and final model explained 23.9% of the variance in total problems. Avoidance coping strategies significantly positively predicted scores at the scale, but not after Bonferroni correction.

Table 5 Significant results of multiple linear regression analyses for ASEBA self-report syndrome scales

ASEBA self-report syndrome scales	Predictors	B	β	p	R ²
1. Anxious/depressed	Age	0.109	0.066	0.630	0.281
	Intellectual disability	2.802	0.136	0.343	
	Self COPE NVI social support	0.039	0.037	0.835	
	Self COPE NVI avoidance strategies	0.518	0.358	0.016*	
	Self COPE NVI problem solving	0.227	0.199	0.287	
2. Aggressive behaviour	Age	0.210	0.225	0.093	0.246
	Intellectual disability	1.677	0.145	0.294	
	Self COPE NVI avoidance strategies	0.276	0.341	0.016**	
3. Internalizing problems	Age	0.276	0.114	0.395	0.300
	Intellectual disability	1.868	0.062	0.657	
	Self COPE NVI social support	0.216	0.140	0.427	
	Self COPE NVI avoidance strategies	0.762	0.364	0.014*	
	Self COPE NVI problem solving	0.235	0.143	0.439	
4. Total problems	Age	0.270	0.125	0.372	0.239
	Intellectual disability	1.363	0.051	0.728	
	Self COPE NVI social support	0.147	0.106	0.561	
	Self COPE NVI avoidance strategies	0.576	0.307	0.043*	
	Self COPE NVI problem solving	0.227	0.154	0.424	

Patients' COPE NVI scores and physical impairments were entered as independent variables on the basis of previous correlation analysis. Age and intellectual disability were included by default as predictors in each regression analysis. $N=51$

ASEBA Achenbach System for Empirically Based Assessment, COPE NVI Coping Orientation to the Problems Experienced—New Italian version

* $p < 0.05$

**Significant p values after Bonferroni correction ($p \leq 0.01$ for model 1, 3 and 4; $p \leq 0.017$ for model 2)

Regressions performed for parent coping

Significant results from multiple linear regression analysis examining the contribution of parental coping strategies on patients' emotional behavioural problems ($N=76$) are presented in Table 6.

The first model explained 24.9% of the variance in withdrawal–depressive symptoms. Avoidance coping strategies of parents and individual intellectual disability add statistically significantly to the prediction of the outcome. Both predictors had positive Beta coefficients and were still significant after Bonferroni correction.

The second model considered somatic problems as dependent variable and overall explained 12.6% of the variance in this scale. Avoidance coping strategies of parents was the only significant predictor of the outcome (positive Beta coefficient) and survived Bonferroni correction.

In the fifth model, considering internalizing problems as dependent variable, the proportion of variance explained was 16.6%. The only significant predictor of the outcome was individual intellectual disability (positive Beta coefficient) which survived Bonferroni correction, whereas avoidance coping strategies of parents showed a trend toward

significance. Predictors of thought problems were examined in the third model, which explained 18.9% of the variance in the dependent variable. Significant predictors of the outcome were individual intellectual disability and parents' social support coping (both with positive Beta coefficient) but they did not survive Bonferroni correction.

Rule-breaking behaviour as dependent variable was examined in the fourth model, which explained 19.4% of the variance in this scale. The variables that significantly contributed to the prediction of scores were cardiomyopathy and problem solving coping strategies of parents, both with negative Beta coefficients that remained significant after Bonferroni correction.

Discussion

Comments to results

In the present study, we considered a large population of young patients with MDs, exploring their cognitive and emotional features, the prevalence of neurodevelopmental

Table 6 Significant results of multiple linear regression analyses for ASEBA parent-report syndrome scales

ASEBA parent-report syndrome scales	Predictors	<i>B</i>	β	<i>p</i>	<i>R</i> ²
1. Withdrawn/depressed	Age	−0.001	−0.001	0.994	0.249
	Cardiomyopathy	−2.934	−0.185	0.155	
	Intellectual disability	5.956	0.317	0.004**	
	Parents' COPE NVI Avoidance strategies	0.458	0.281	0.009**	
2. Somatic complaints	Age	0.030	0.037	0.742	0.126
	Intellectual disability	1.255	0.083	0.486	
	Parents' COPE NVI Avoidance strategies	0.383	0.342	0.003**	
3. Thought problems	Age	0.024	0.037	0.749	0.189
	Intellectual disability	2.778	0.256	0.028*	
	Parents' COPE NVI Social support	0.172	0.269	0.021*	
	Parents' COPE NVI Avoidance strategies	0.176	0.187	0.102	
4. Rule-breaking behaviour	Age	0.035	0.100	0.475	0.194
	Intellectual disability	0.417	0.073	0.525	
	Cardiomyopathy	−1.758	−0.363	0.011**	
	Parents' COPE NVI Problem solving	−0.122	−0.314	0.007**	
5. Internalizing problems	Age	0.347	0.253	0.066	0.166
	Intellectual disability	6.816	0.304	0.008*	
	Cardiomyopathy	−3.204	−0.169	0.216	
	Parents' COPE NVI Avoidance strategies	0.383	0.197	0.079	

Patient's physical impairments and parents' COPE NVI scores were entered as independent variables on the basis of previous correlation analysis. Age and intellectual disability were included by default as predictors in each regression analysis. $N=76$

* $p<0.05$

**Significant p values after Bonferroni correction ($p\leq 0.013$ for model 1, 3, 4 and 5; $p\leq 0.017$ for model 2)

ASEBA Achenbach System for Empirically Based Assessment, COPE NVI Coping Orientation to the Problems Experienced—New Italian version

and psychiatric disorders and both their own and their parents coping styles. Furthermore, we assessed the role of individual (age, physical impairment, ID, coping styles) and familial factors (SES, parental coping styles) on psychological outcomes of patients. We used well-validated instruments to collect data, and patients included in the study were all genetically defined.

Our data confirm previous literature and our hypothesis regarding neurodevelopmental disorders such as ID and ASD among patients with DMD: these conditions were found only in the DMD subsample, resulting in 22.3% of the total sample for ID, and 6.25% of the total sample for ASD, which are high prevalences if compared to general population [64, 65].

In relation to emotional/behavioural problems, as we hypothesized, the first-step screening instruments returned a high rate of internalizing problems, both among young adults (18%) and—at a major extent—among children (24.6%) according to ASEBA questionnaires. The overall prevalence of internalizing problems in children resembles that found in our previous study (23.4%) [15] and in Ricotti and colleagues' study (24%) [16], which considered a sample of children with DMD similar to our in terms of size and used the same questionnaire. Even exploring SDQ profiles, internalizing problems area (comprising emotional and peer problems scales) was still the highest, and, among children, it was drawn by a serious presence (17.54%) of problems with peer relations, as compared to general population of western European countries reported in previous studies [61–63]. These data suggest that children with MDs experience symptoms of anxiety and depression that might be associated with difficulties in gaining peer acceptance and in establishing friendships. In contrast to the aforementioned study by Ricotti and colleagues', we found a very low prevalence of externalizing problems (1.64% among children and 0% among young adults, according to ASEBA).

When we observed results of the second-step in-depth diagnostic assessment, we noticed that the percentage of patients meeting clinical caseness is considerably reduced: rates of ADHD, internalizing disorders and conduct/oppositional disorders are in line with prevalence rates reported in general population studies, both for children and for adults (see [66–68] for ADHD; see [60, 69] for internalizing disorders; see [60] for conduct/oppositional disorders). Nevertheless, in children with BMD, we found a much higher rate of ADHD (33%) compared to children with DMD (1.9%), LGMD (0%) or general population data [66]. This observed frequency of ADHD in BMD is similar to that reported by two previous studies [14, 18], where a notably higher percentage compared to that observed in DMD has been also replicated [14].

Moreover, comparing the three different types of MDs on overall frequency of emotional/behavioural disorders, BMD subsample was that with the highest rate (23.53%).

Since prevalence of psychopathology in our overall sample was found to be aligned with that of general population, our interest was directed at the exploration of coping styles of parents and patients, to verify whether they adopt strategies to cope with stressful situations that are protective against psychological distress. Compared to general community, caregivers of patients with MDs relied less on avoidance, problem solving and social support, while they had a higher level of positive attitude. Lower problem solving was also evident in coping profile of patients, together with a lower tendency to turn to religion. These results are in line with our expectations of a greater use of secondary control coping (positive attitude) over primary control coping (problem solving) compared to general population. In previous research on chronic illness, less avoidance attitude and higher acceptance have been related to better psychological adjustment [41, 46]. Based on this knowledge, our observed parental coping profile might be considered adaptive. In a transactional stress and coping model perspective, parents who cope with their children's condition accepting the reality—rather than denying it—and positively reinterpreting the experience, (e.g., through the finding of opportunities for growth), could have a better emotional regulation in the long term, with a positive influence on children's emotional well being.

The lower use of primary control coping (i.e., social support by parents and problem-focusing by both parents and children compared to general population) could lead to different interpretations. Social support could be less used because of family social isolation and stigma. Alternatively, it could be interpreted as a way to adapt; in fact, looking at research evidences on coping with not easily controllable stressors, a high use of coping strategies aimed at directly change the source of stress or the emotional reaction to it (as problem solving and seeking social support), was found to be related to a worse emotional/behavioural adjustment compared to strategies aimed at change aspects of oneself to accommodate to the stressor (such as positive attitude) [41, 61, 62]. These findings do not deny the role of psychological support from the social network in reducing the effect of stress, which can be considered, as described by several studies, a buffer between coping with an event and stress [30], but it might be not sufficient on its own to guarantee adjustment [61, 62]. This consideration can be also valid for religious coping, which has not been found to be strongly related to psychological well being [62].

Based on these data, we were further interested to test the association between coping styles and psychological outcomes in our sample, taking also into account individual variables. The results of the regression analyses consistently

confirm our hypotheses, showing the use of avoidant coping strategies to be associated with higher risk of emotional/behavioural problems in patients with MDs. A higher risk of psychological distress was observed in patients with MDs both when avoidant coping was used by their parents and when they were themselves to use it. These findings support the aforementioned past study of Thompson and colleagues [34], which indicated higher parent use of disengagement coping to be associated with poor adjustment in children with DMD, as well as previous studies on chronically ill patients where their self-reported use of avoidance was consistently found to be associated with psychological distress, especially anxiety and depression [40]. In our sample, parents who were more prone to use avoidant coping were more likely to have children with increased levels of internalizing problems. On the other hand, if were patients themselves to make a greater use of avoidant coping strategies, they were more likely to report higher levels of both internalizing (i.e., anxiety, depression) and externalizing problems (i.e., aggressive behaviour).

We also noticed that higher use of social support from parents was associated with an increase in children's thought problems, supporting the hypothesis that this coping strategy may not always be useful, as it could reflect a tendency to vent emotions, which is indeed a form of disengagement [58]. However, caution is needed in interpreting this result, because after Bonferroni correction, the *p* value was far from significant level. On the other hand, we found less rule-breaking problems in patients whose parents used more problem-focused coping, suggesting a possible protective role of this strategy, especially when it is used for the sources of stress perceived as more controllable, such as problems related to school or home help or providing adaptive equipment. No such association was found for problem solving when used by patients, in line with mixed findings regarding this strategy reported in literature [40, 58].

Observing parents' rating of their childrens' psychological difficulties, regression analyses also showed that patients' intellectual level was related to their emotional/behavioural outcome: the presence of comorbid ID was in fact related with increased internalizing problems and thought problems. This is consistent with numerous studies indicating ID as a risk factor for developing psychopathology [70, 71].

Finally, it is interesting to note that measures of disease progression (age and wheelchair use) did not contribute significantly to reported elevated behaviour problems, confirming previous evidences suggesting coping style and adjustment as more salient to psychological outcome than disease severity [72, 73].

Strengths and weaknesses of the study

This study adds to current knowledge regarding neurodevelopmental disorders and psychological status among people with MDs. It also provides new knowledge on how families cope with stress and on the relationship that specific coping strategies and clinical aspects of MDs have with the risk of psychological distress. We consider strengths of our study the use of a genetically well-defined clinical population and of validated instruments directly administered to both patients and their parents.

However, results should be interpreted with some limitations in mind.

First, even though this sample is appropriate for a mono-centric study, our three subgroups of MDs were not homogeneous in terms of size (the majority had DMD), whereas to perform more precise group comparisons, larger and more balanced sample sizes will be needed. This limitation also concern comparisons of coping strategies between parents and general population or patients and general population.

Moreover, since the COPE questionnaire is not validated for children, we needed to exclude them from the investigation of patients' self-reported coping styles. In future studies, it will be desirable to address this limitation.

In the second-step, in-depth diagnostic assessment, additional structured interviews (i.e., ADOS and DAWBA) were administered only in a subsample of 38 patients (34.2% of the total sample), according to our assessment procedure: this may have led to an underestimation of the true rate of psychopathology in this special clinical population. Nevertheless, the utility of behavioural screening instruments for risk stratification has been largely demonstrated [74, 75]. Sheldrick and colleagues [75] reported that children who screened positive at CBCL 6–18 and SDQ were approximately 2–5 times more likely to qualify for a psychiatric diagnosis compared to those children who screened negative.

Since the study has a cross-sectional design, causality cannot be fully investigated and interpretation of results needs caution.

Finally, the large number of statistical tests performed on the same data set may have increased the probability of statistically significant results by chance. To control this risk, we have applied Bonferroni correction to multiple analyses.

Future research

Future research should use larger and more balanced sample sizes to draw more definite conclusions and to perform more precise group comparisons. In particular, it would be interesting to extend BMD sample to better explore its neurodevelopmental and emotional profile, since there are suggestions of a greater vulnerability to psychopathology

among BMD patients. Moreover, designing and conducting longitudinal studies can improve understanding the role of parental coping later in emotional/behavioural development of children. Finally, it would be interesting to explore coping strategies in younger children, with less than 16 years. To do this, it will be desirable to administer instruments for the study of coping specifically designed for children and with Italian normative data.

Conclusions

In conclusion, our study indicates that, despite the relatively high presence of neurodevelopmental disorders (i.e., ASD, ID, ADHD) and internalizing symptoms, children and young adults with MDs exhibit rates of diagnosed psychopathology that are in line with those of general population. Within families of our sample, parents face stress through the use of a coping style known to be more adaptive in the context of a chronic illness, wherein positive reinterpretation and growth and acceptance prevail over denial and behavioural or mental disengagement. Findings of this research also show a link between coping strategies and psychological adjustment in patients with MDs. The most consistent relationship was with avoidance, that, when used by parents, predicts internalizing symptoms in patients, and, when used by patients themselves, predicts both internalizing and externalizing symptoms. Moreover, the comorbid presence of ID is indicated as an additional risk factor associated with worse emotional and behavioural outcomes, while disease progression is not. Given the high prevalence of internalizing symptomatology reported in children and young adults with MDs, results of this study suggest that interventions targeting both parents and patients, aimed at discouraging use of avoidant coping and, on the other hand, promoting use of adaptive coping (i.e., positive reinterpretation, acceptance, problem solving for situations that are controllable) could reduce—via indirect and direct mechanisms—anxiety and depression and, more generally, psychological distress among patients.

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Compliance with ethical standards

Conflicts of interest The authors declare that they have no conflict of interest.

Ethical standards The study was approved by the Ethical Committee of the Scientific Institute IRCCS Eugenio Medea according to the Declaration of Helsinki.

Informed consent All patients provided written informed consent for participation in the study.

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