



Original article

Social involvement issues in patients with Becker muscular dystrophy: A questionnaire survey of subjects from a patient registry

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Abstract

Background: Little is known about the relationship between Becker Muscular Dystrophy (BMD) and developmental problems, school life, employment, and mental problems. We aimed to clarify whether BMD is a risk factor for developmental disorders, problematic behavior, psychiatric diseases, and other social difficulties in school life and employment.

Methods: Adults with genetically or immunohistochemically confirmed BMD from the Registry of Muscular Dystrophy in Japan (REMUDY) were asked to complete a questionnaire regarding patient history, school life, employment, and mental problems.

Results: In total, 125 (68.3%) of 183 participants with BMD (median age, 37.2 years) completed the questionnaire. Of these, ten had developmental disorders (mental retardation, autism, and speech disturbance). Fifty-eight (44%) experienced bullying in school, and 39 felt the reason for bullying was physical handicap. Sixteen participants experienced problematic behavior such as cutting class, domestic violence, violent incidents, suicide attempts, or self-mutilation. Employment histories were noted by 92 (73%), of whom 15 could not continue to work due to physical handicaps. Fifteen participants had psychiatric disorders, with 5, 3 and 1 having neurosis, depression, and bipolar disorder, respectively. The other 6 participants with psychiatric disorders did not specify their diagnoses. Patients carrying a Dp140 expression change had significantly more incidences of developmental disorders, but not bullying, problematic behavior, workplace difficulties, or psychiatric disorders.

Abbreviations: BMD, Becker muscular dystrophy; DMD, Duchenne muscular dystrophy; Remudy, Registry of muscular dystrophy in Japan; NCNP, National Center of Neurology and Psychiatry; SD, standard deviation

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Conclusions: Patients with BMD risk bullying and workplace difficulties, as well as developing psychiatric disorders. Parents, teachers, and supporters should be mindful of the daily environment of BMD patients and provide support to help them cope with stress.

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Keywords: Becker muscular dystrophy; Dystrophinopathy; Patient registry; Questionnaire; Psychiatric diseases; Education; Bullying; Employment; Mental problems

1. Background

Becker muscular dystrophy (BMD, OMIM 300376) is an X-linked recessive form of muscular dystrophy caused by mutations in the dystrophin gene (*DMD*) on chromosome Xp21.2 [1]. The *DMD* gene is the largest gene identified in humans and contains 79 exons. Mutations in the gene result in a deficiency of normal dystrophin protein [2]. BMD is typically associated with mutations that maintain the open reading frame, producing an internally altered but partially functional dystrophin protein, with an intact C terminal domain [3,4]. Patients with BMD have variable phenotypes, but are less severely affected than those with Duchenne muscular dystrophy (DMD), and have a much longer normal life expectancy [4].

Neurodevelopmental disorders are increasingly recognized as characteristics of DMD [5,6]. For instance, cognitive impairment, autism, hyperactivity, and intention disorders are frequently found in patients with DMD [5]. Related mental problems such as depression and neurosis have also been reported [6–8]. In contrast, little is known about whether developmental disorders are associated with BMD and the mental health state of patients with BMD, although some cognitive issues experienced by patients with BMD appear to be similar to those experienced by patients with DMD [9]. It is often the case that patients with BMD experienced certain mental problems and difficulties in daily life (e.g., bullying) back when they were attending school. Moreover, some patients also experience mental stress at the workplace, which can lead to depression and neurosis.

Given that patients with BMD commonly develop depression and other psychiatric disorders, physicians should diligently monitor their mental health to detect early signs that would allow for early therapeutic intervention. Indeed, their mental problems potentially are derived from their surrounding environment and mental stress; in particular, being physically handicapped amidst healthy classmates in regular school classes. Against this backdrop, this study aimed to analyze whether BMD is a risk factor for psychiatric disorders, and if so, to analyze factors that contribute to psychological dysfunction in patients with BMD and other dys-

trophinopathies. To clarify the potential relationship of BMD with social difficulties and psychiatric disorders, mental/developmental problems, and growth environment, we performed a questionnaire survey of subjects from a patient registry with confirmed BMD.

2. Methods

2.1. Aim and design

This study aimed to clarify whether BMD is a risk factor for developmental disorders, problematic behavior, and/or psychiatric diseases. We also assessed other social involvement difficulties of patients with BMD in their educational, developmental, and working environments.

2.2. Patient registry, organization, registration method, data collection, and ethical approval

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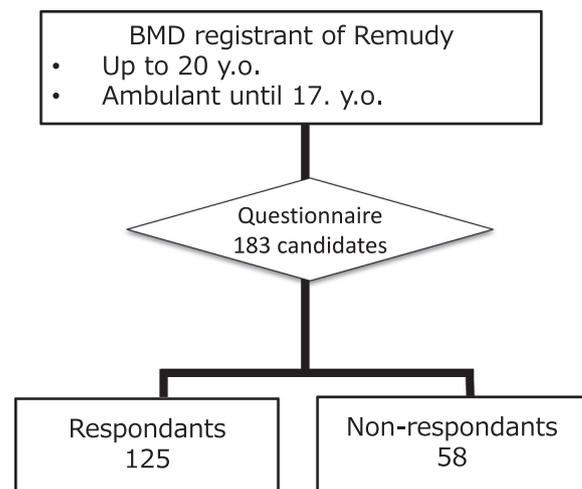


Fig. 1. Patient recruitment. We sent questionnaire forms to 183 registrants with dystrophinopathy who were aged ≥ 20 years and were ambulant at age 17. Of these, 125 responded.

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