Juvenile fibromyalgia syndrome. Interdisciplinary treatment

Młodzieńczy zespół fibromialgii. Leczenie interdyscyplinarne

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Summary
Fibromyalgia syndrome (FM) belongs to soft tissue pain syndromes of unknown cause, also referred to as "soft tissue rheumatism". It is characterized by chronic widespread pain as well as additional symptoms such as fatigue, sleep and mood disturbance and cognitive problems. There is more and more data showing that this condition may start at a young age or even in childhood, adversely affecting development processes and resulting in dysfunctional social and family relationships. Because of the multifaceted character of fibromyalgia the efficient treatment of this disorder can be difficult and requires comprehensive care.

This work reviews most recommended procedures used in integrated treatment programmes for juvenile fibromyalgia syndrome (JFM).

Introduction
Fibromyalgia is a non-inflammatory syndrome characterized by chronic and widespread musculoskeletal pain. In 2010 ACR (American College of Rheumatology) suggested new diagnostic criteria that do not include a tender point test but include widespread pain index (WPI) and symptom severity (SS) scale. The WPI includes a list of 19 areas in which a patient has had pain over the last week. Each painful location is associated with one point, and then raw data should be added. On the other hand, the symptom severity scale measures the intensity of such symptoms as fatigue, sleep problems (difficulty falling asleep, nighttime waking) and cognitive disturbances. The value of 0 to 3 has to be selected on a numerical scale. If a patient has additional symptoms like irritable bowel syndrome, depression, headache it is necessary to add 3 points to the general score. Fibromyalgia is diagnosed when the WPI score is at least 7 and the SS score is at least 5, or the WPI score is at least 3 and the SS score is at least 9 (Table I).

In the past, Yunus and Masi suggested classification criteria for juvenile fibromyalgia syndrome (Table II). As in the case of adults, JFM is associated with persistent and widespread musculoskeletal pain lasting for at least 3 months and located in at least 3 body areas. Additionally, during a palpation examination a patient has to feel tenderness on compression in at least 5 out of 18 tender points located in specific anatomic areas (located at the sites of tendon attachments or along their courses). Moreover, additional criteria have to be met and they include fatigue, sleep and mood disturbances, chronic headache, irritable bowel syndrome etc. In order to diagnose JFM a patient has to meet all conditions from...
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Table I. Fibromyalgia diagnostic criteria ACR 2010 [1]

| A patient satisfies diagnostic criteria for FM if the following 3 conditions are met: |
| 1. Widespread pain index (WPI) ≥ 7 and symptom severity (SS) scale score ≥ 5 or WPI 3–6 and SS scale score ≥ 9 |
| 2. Symptoms have been present at a similar level for at least 3 months |
| 3. The patient does not have a disorder that would otherwise explain the pain |

Ascertainment:

1. WPI: note the number areas in which the patient has had pain over the last week. Score should be between 0 and 19
   - shoulder girdle, left
   - hip (buttock, trochanter), left
   - shoulder girdle, right
   - hip (buttock, trochanter), right
   - lower arm, left
   - lower leg, left
   - lower arm, right
   - lower leg, right
   - upper arm, left
   - upper leg, left
   - upper arm, right
   - upper leg, right
   - jaw, left
   - upper back
   - chest
   - abdomen
   - neck

2. SS scale score:
   - Fatigue
   - Unrefreshing sleep (difficulty falling asleep, nighttime waking)
   - Cognitive disturbances
   For each of the 3 symptoms above, indicate the level of severity over the past week using the following scale:
   - 0 – no problem
   - 1 – slight or mild problems, generally mild or intermittent
   - 2 – moderate, considerable problems, often present and/or at a moderate level
   - 3 – severe, pervasive, continuous, life-disturbing problems
   Considering somatic symptoms in general, indicate whether the patient has:
   - 0 – no symptoms
   - 1 – few symptoms
   - 2 – a moderate number of symptoms
   - 3 – a great deal of symptoms
   The SS scale score is the sum of the severity of 3 symptoms (fatigue, sleep disturbances, cognitive symptoms) plus the extent of somatic symptoms in general. The final score is between 0 and 12

Somatic symptoms that should be considered: muscle pain, irritable bowel syndrome, fatigue/tiredness, thinking or remembering problems, muscle weakness, headache, abdominal pain, tingling, numbness, dizziness, insomnia, depression, constipation, pain in the lower abdomen, nervousness, nausea, chest pain, vision disturbances, diarrhea, dry mouth, wheezing, itching, Raynaud’s phenomenon, hives, ringing in ears, muscle weakness, headache, abdominal pain, tingling, numbness, dizziness, insomnia, depression, constipation, Raynaud’s phenomenon, hives, ringing in ears, muscle weakness, headache, abdominal pain, tingling, numbness, dizziness, insomnia, depression, constipation, Raynaud’s phenomenon

Table II. Diagnostic criteria for juvenile primary fibromyalgia syndrome [2]

| I. Main criteria |
| 1. Widespread musculoskeletal pain located in at least three areas for at least 3 months |
| 2. No underlying disease that might explain symptoms |
| 3. Laboratory results within the norm or negative |
| 4. At least 5 typical tender points |

| II. Additional criteria |
| 1. Sleep problems (difficulty falling asleep, nighttime waking) |
| 2. Chronic anxiety and/or tension |
| 3. Irritable bowel syndrome |
| 4. Chronic headache |
| 5. Subjective soft tissue swelling |
| 6. Pain modulated by physical activity |
| 7. Pain modulated by anxiety and/or stress |
| 8. Pain modulated by weather |

In order to diagnose JFM 4 main criteria and 3 additional criteria have to be met or first 3 main criteria, 4 tender points and 5 additional criteria have to be met
abnormal transmission of pain stimuli within the central nervous system (CNS) [3]. Moreover, more and more data indicate that genetic and environmental factors work together in the pathogenesis of FM [4].

FM is estimated to develop in 1.2–6.2% of the developmental-age population [5–7], whereas the incidence of JFM in rheumatology clinics ranges between 7% and 35% [8–10]. This syndrome is more frequently diagnosed in adolescent girls than in boys, especially in highly controlling families where there are no flexible educational patterns [2, 5–7, 11]. Due to heterogeneity of symptoms responsible for a diagnosis a diagnostic process is sometimes prolonged, and the treatment itself is usually symptomatic [12]. Additionally, in practice we should consider coexistence of fibromyalgia symptoms with, for example, juvenile arthritis or chronic fatigue syndrome [9, 13]. Moreover, estimated data confirm a strong relationship between JFM and mental disorders. In mental health hospitals the incidence of symptoms is even higher than in the general population [13, 14].

There are ambiguous data on the prognosis of FM in a pediatric population. However, it seems that the presence of FM symptoms during childhood is a poor prognostic factor [15]. Recently, researchers have attempted to determine how methods to treat FM affect clinical symptoms of this syndrome. In order to do that statistical analyses based on a rheumatologic register of patients with FM at the age of 9–21 years have been performed. It has turned out that the incidence of symptoms (sleep problems and headache, especially) increased irrespectively of treatment. Nonetheless, the most recommended interventions included physical therapy sessions, psychiatric consultations, biofeedback training and pharmacotherapy [16]. The study results also suggest there are no differences with regard to a diagnostic process and efficacy of therapy applied based on a criterion of the patient’s age. However, as it has been observed by Eraso et al. [17], children below 10 years demonstrate higher intensity of symptoms belonging to the FM spectrum compared to a group of patients who are older. On the other hand, Weiss [18] notes it is necessary to consider a child’s sex in a diagnostic process. Although the diagnosis of FM mainly regards adolescents, the authors emphasize that a male sex predisposes to more serious functional problems and poorer adaptation to symptoms of this syndrome.

The need to introduce an intervention rapidly is also supported by the fact that children and adolescents with JFM exhibit more disturbances with regard to physical and psychosocial functioning compared to adolescents with cancer or other rheumatic disease [19–21]. Moreover, the diagnosis of FM is often accompanied by the presence of anxiety and cognitive disturbances [22]. One of studies demonstrated that more than 70% of adolescents with suspected JFM had at least one psychiatric diagnosis, and anxiety disorders (specific and social phobias, generalized fear and panic attacks) were diagnosed the most frequently. Furthermore, 19% of patients met the ADHD criteria (a subtype with predominance of attention concentration deficits) [23]. Adolescents with FM also have a specific pattern of psychic constitution, and they also function within families with a disturbed organisational structure where excessive criticism and high expectations of parents are predominant [13, 20]. As a consequence, it may lead to inhibited development of effective adaptation strategies that are necessary in everyday life and may slow down the healing process.

The article aims to review current information on interventions that are the most commonly used in the treatment of juvenile fibromyalgia syndrome.

### Interventions targeted at physical activity

More and more evidence supports the effectiveness of regular physical exercises in the rehabilitation of patients with FM [23]. Imaging tests with adults suggest that physical activity plays a regulatory role in processes of pain modulation and perception [24]. The introduction of moderate effort, especially in a form of aerobic training, seems to have long-term benefits also in adolescents. Based on the results collected by Stephens et al. [25] 12-week aerobic training of moderate intensity turned out to be a safe form of intervention, namely it did not result in disease symptom exacerbation. Additionally, it positively affected reduction of fatigue and pain, and increased physical capability of participants of such training sessions. However, reports show that adolescents dedicate a relatively low amount of time for moderate or intensive physical effort [26]. Additionally, despite the fact that it has been confirmed that interventions targeted at physical activity are effective, the demand for physical activity reduces with age [27]. It also corresponds with the results of an actigraphic study that demonstrated reduced locomotor activity in a group of older children compared to younger patients [26]. Nonetheless, it seems that effects of physical training are especially beneficial as it eliminates a subjective component of pain and other symptoms that are associated with JFM. For example, Olsen [28] et al. have demonstrated that interventions aimed at regular effort reduce a subjective assessment of pain, eliminate signs of fatigue and reduce difficulty falling asleep. Due to increased likelihood of traumas and overloading in the population suffering from JFM specialists emphasize it is necessary to introduce individualized training programs so as to optimize the effect of pain elimination.
during and after effort [25, 29]. Moreover, it may also be beneficial to adjust exercise types to patient’s age and maintain motivation for training. In one of their studies Reid et al. [21] have demonstrated that when a child is encouraged to maintain activity despite mild pain they endure the activity better, and pain tolerance is improved and it is not observed when negative statements are present, as then a child stops their effort.

**Pharmacological interventions**

Pharmacotherapy plays a relatively small role in the treatment of JFM symptoms. In the population of adolescents large multicenter clinical trials regarding mechanisms of actions and side effects of treatment are missing [18, 29]. At the beginning of the 90s Romanò [30] observed a group of 15 patients with JFM for 2 years and demonstrated good therapeutic effects of cyclobenzaprine in 11 patients. However, when analyzing pharmacotherapy that is used the most often, it seems that standard treatment tends to include treatment combined with low doses of tricyclic antidepressants (TCA) and non-steroid anti-inflammatory drugs [22]. Pharmacotherapy also includes antiepileptic drugs, such as gabapentin, and its use has been recently higher in adolescent males [18]. Higher consumption of medicinal products can also be observed in young adults and according to some sources it may be related to continued or exacerbated symptoms of FM, as well as with a better access to reimbursed medicinal products in case of adults. It should also be mentioned that a demand for supplements such as vitamins and herbal products increases with age [27].

**Cognitive-behavioral interventions**

Cognitive-behavioral interventions belong to therapeutic methods that are the most frequently monitored under empiric conditions. Reviews of randomized trials indicate their efficacy with regard to pain elimination among children and adolescents with chronic as well as recurrent pain [31]. Cognitive-behavioral therapy (CBT) is aimed to modify erroneous thought patterns and to replace them with strategies that are indicated for better adaptation. In a population of patients with FM similar interventions are used to enhance control over pain and clinical symptoms of this syndrome [32].

Many studies confirm the efficacy of similar procedures. According to Kashikar-Zuck et al. [33] 16-week sessions provide better pain control and a lower level of disability as well as fewer depressive symptoms compared to baseline values. Degotardi et al. [34] have also obtained partial remission of symptoms in a group of 67 patients as a result of using CBT with components of autorelaxation. During an 8-week intervention authors observed significant reduction of pain, somatic symptoms, anxiety and fatigue as well as improved sleep quality immediately after therapy.

Educational sessions are often used as an alternative or supplementation of cognitive-behavioral therapy. They are dedicated to general problems associated with specific features of the syndrome. Contrary to CBT they emphasize only an informative aspect, and they do not model adaptive changes in individual functioning. Despite a simplified approach it has recently been proven that even under conditions of urgent psychiatric hospitalization psychoeducation classes combined with relaxation techniques provide benefits such as reduced levels of stress, muscle tension or indigestion. Additionally, they improve the concentration level, increase self confidence and patient’s mental comfort [35–37]. However, when comparing the efficacy of educational therapy with CBT investigators found some differences. Thanks to educational sessions with elements of muscle relaxation instructions adolescents experienced significant improvement with regard to their functional disability and symptoms of depression. However, a group participating in cognitive-behavioral therapy demonstrated better functioning after the end of the active treatment phase and during a 6-month follow-up [35]. In their last study Sil et al. [37] attempted to identify factors that might affect functional disability of children. Adolescents that have already participated in educational classes and CBT have been asked to participate. The analysis has demonstrated that effective coping with disease (i.e. self-efficacy level, higher self-assessment) was a good prognostic factor with regard to symptoms of functional disability. This effect was especially observed in the CBT group. On the other hand, other variables, like pain intensity or depression symptoms did not affect the prognosis of studied symptoms.

**Summary**

Guidelines of experts on the management of juvenile fibromyalgia tend to recommend non-pharmacological strategies that are only partially supported by pharmacotherapy [18, 29]. However, due to a multifactorial nature of this syndrome it is difficult to make a rapid diagnosis and to introduce treatment that is mainly aimed to reduce vegetative symptoms (musculoskeletal pain, long-term sleep disturbances and fatigue). Currently, it is recommended to combine several therapies in interventional programs so as to affect deficits associated with FM symptoms parallelly. It also has to be emphasized that the majority of interventions used has been modified for the needs of younger patients based on strategies used in the treatment of adults.
and their efficacy has not been sufficiently verified in controlled clinical trials. Nonetheless, it seems that when physical activity starts to be treated as a habit, relatively good effects are observed [25]. However, with regard to the efficacy of cognitive-behavioral interventions it has recently been suggested that they may regard more global mental changes regarding a cognitive assessment of pain or perception of one’s own efficacy of pain management [32]. Future studies should be focused on the identification of homogenous subgroups based on a holistic model of health that as a result may fill in blanks to understanding JFM pathogenesis, and in the long-term it may result in a lower incidence of FM among adults.

Reference

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