

The importance of psychosocial factors in management of pulmonary arterial hypertension patients

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INTRODUCTION

In recent years, an approach to treating pulmonary arterial hypertension (PAH) patients, which emphasises their quality of life, has been dynamically developing [1–3]. It is important for the therapeutic process both to strive to prolong the patients' lives and to improve their everyday functioning, with a clear focus on restoring their usual personal and family life profile [4]. Even though the development of new PAH therapies extends the life expectancy of PAH patients, a fully curative therapy, especially without serious side-effects following acute and chronic drug administration, is still unavailable [5]. The patient's life is prolonged but symptoms typical of PAH remain, so patients need to learn how to cope with their uncertain future and complex treatments which, more often than not, lead to side effects changing a person's lifestyle [6]. More frequent hospitalisations, limited physical and everyday life activity, increasing dependence on others, less frequent social interactions, impossibility to take certain life and social roles, loss of professional position, worse financial situation, and the necessity to adapt the current lifestyle to the disease may all contribute to the emotional functioning of patients with PAH [6]. Another very important psychological problem for PAH patients is the fact that those among them qualified for the National Transplant Waiting List need to cope with the stress connected with waiting for an organ, expectations of complex surgery, and the subsequent rehabilitation.

The latest guidelines regarding pulmonary hypertension diagnosis and treatment, which were issued in 2015, emphasise that the medical team taking care of this group of patients ought to have knowledge and skills in the area of the impact of the disease on the psychological, social, emotional, and spiritual functioning of patients and their families; the

team should also cooperate closely with specialists, including a psychiatrist, a clinical psychologist, and a social worker, in order to provide help for patients with serious problems [7]. Most commonly, however, it is the physician who is most important to a patient, and who should coordinate the entire therapeutic programme. Therefore, knowledge of psychological, psychiatric, or existential factors connected with PAH is indispensable [8].

'INVISIBLE DISEASE', DEPRESSION, ANXIETY DISORDERS

The adjustment to interpersonal and social issues unique to the symptoms and treatment of their illness constitutes a hardship for many individuals diagnosed with PAH. Although a typical patient with PAH diagnosis experiences some degree of physical limitation, either requiring frequent resting or resulting in significant exercise intolerance, clinical observation would indicate that, at first glance, only a few of them 'appear sick'. This can create a false perception that the individual is 'not very sick' or that they might be exaggerating in order to win sympathy or other indirect gains, such as being relieved from family, work, or social role responsibilities. These issues can be difficult to cope with in the early stages of the disease, particularly if patients themselves have not fully accepted that they are ill. Such struggles frequently lead to reduced social functioning. Social interactions are important for gaining and maintaining social acceptance, but for many individuals with a PAH diagnosis, physical limitations can lead to increased social isolation [9].

Most specialists taking care of PAH patients frequently observe serious psychological problems (depression, anxiety disorders, or panic attacks). A common problem reported by

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PAH patients is the feeling of social isolation, as mentioned above, and the fact that their friends and family show little understanding or knowledge of their disease. Among the PAH patients, 80.4% reported that their friends, family, and colleagues do not understand PAH [10]. The feeling of social isolation is especially noticeable in patients under 30 years old, i.e. in the age group where social interactions may constitute a particularly important part of everyday life. It is observed that PAH has an influence on other aspects of mental health as well. In a study of 101 PAH patients, almost half (48%) suffered from mild or severe anxiety; approximately 1/3 experienced depressive symptoms (32.6%) or stress (27.6%) [11]. A study designed by Löwe et al. [12] aiming at defining the prevalence of emotional imbalance in PAH patients (70.1% of female patients; average age 47.8 ± 12.7 years) showed that 35% of patients suffered from mental disorders, and the most frequently diagnosed ones were serious depression (15.9%) and panic attacks (10.4%). In another study, conducted on a larger group of 100 patients, depression was diagnosed in 55% of PAH patients, whereas there were severe depressive disorders in 15% of patients, and mild or moderate depression was diagnosed in 40% of patients [13]. It has been observed that the frequency of mental disorders in PAH patients significantly increased as their level of activity deteriorated. Emotional disorders were diagnosed in 17.7% of patients in New York Heart Association (NYHA) class I, whereas in the case of NYHA class IV patients — in 61.9% of them [12]. A noteworthy limitation of this study is the method of selecting the sample group, as the study was carried out on patients who belonged to an association of PAH patients. Such patients may not be representative of the emotional and social functioning of their population because they already belong to an organisation that helps them to adapt to living with a disease and because they have already developed active strategies of coping with their situation. It may be assumed, therefore, that the rate of anxiety disorders and depression in this group is lower than the entire PAH patient population. Nevertheless, a prevalence of mental disorders at a level of 62% is incredibly high taking into consideration the social support and active strategies of coping with the situation, which result from joining a support organisation [12]. As a consequence, it is unclear what ratio applies to the population of patients who do not have access to such sources of social support. Although PAH patients frequently suffer from anxiety and depression, only some of them undergo appropriate treatment. In the analysed study, only 24.1% of PAH patients suffering from mental disorders were referred for psychopharmacological and psychotherapeutic treatment [12].

Apart from the emotional support and knowledge provided by a doctor, every PAH patient may benefit from different forms of psychological and psychiatric actions. First reports indicate that cognitive-behavioural therapy and psychotherapy groups bring positive results, especially if

psychotherapy is combined with psychopharmacological therapy [14]. An interesting study by Shah et al. [15] suggested a potential connection between lower mortality rate and the development of PAH in adult patients using selective serotonin re-uptake inhibitors; however, further studies are required in this area.

QUALITY OF LIFE

Overall, the debilitating impact of PAH on health-related quality of life (HRQoL) is clear when compared with other medical conditions of severity similar to illnesses such as chronic obstructive pulmonary disease and renal failure, and those conditions are well recognised as being severely debilitating and life-threatening, e.g. spinal cord injury, interstitial lung disease, or treatment-resistant cancer [16]. An interesting comparison of quality of life (QoL) in PAH and chronic thromboembolic pulmonary hypertension (CTEPH) patients was presented by Pfeuffer et al. [17]. CTEPH is a rare, progressive pulmonary vascular disease, that is usually a consequence of prior acute pulmonary embolism [18]. CTEPH usually begins with persistent obstruction of large and/or middle-sized pulmonary arteries by organised thrombi [19]. Although HRQoL was reduced in both cohorts of patients, individuals diagnosed with CTEPH scored lower in nearly all Short Form 36 parameters, including significantly worse metrics for “mental health” ($p = 0.01$) and “mental component summary score” ($p = 0.02$). Depression was also more frequent in patients with CTEPH (56%) than in patients with PAH (30%, $p = 0.03$). Overall, depression and anxiety correlated with most SF-36 scales in both PAH and CTEPH. In CTEPH, depression also correlated with the Borg Dyspnoea Scale ($r = 0.44$, $p = 0.01$). These patients also had significantly lower $p\text{CO}_2$ levels compared with the PAH cohort, which reflects a more severe ventilation/perfusion mismatch. All other haemodynamic and functional parameters did not differ across the groups. While both cohorts of patients suffered from a reduced HRQoL as well as depression and anxiety, decreases in mental health parameters were more pronounced in the CTEPH cohort. This suggests the need for intensive efforts to improve early detection, especially in dyspnoeic patients with classical risk factors for PAH and CTEPH and mental health interventions alongside routine clinical care provided to patients diagnosed with PAH or CTEPH [17].

DIAGNOSTIC QUESTIONNAIRES

In order to initiate appropriate psychological care, a diagnosis is needed. It seems that in the case of PAH the use of screening surveys, i.e. questionnaires allowing identification of patients who urgently need psychological or psychiatric assistance, would be a good solution [12, 14]. David Goldberg’s General Health Questionnaire deserves recommendation. Two versions of this questionnaire are available in Poland — the short, called GHQ-12, and a version with sub-scales assessing four categories of symptoms (somatic symptoms, anxiety and

insomnia, social dysfunction and severe depression), called GHQ-28 [20].

Screening for depression in the PAH population remains challenging, because depressive symptoms, such as low energy or fatigue, can mimic the symptoms of the underlying disease state. Therefore, they can sometimes be ignored [21]. The Beck Depression Inventory may be used for depression screening. The inventory comprises 21 questions. Four variants of answers are available, reflecting an increasing severity of symptoms [22]. The Hamilton Anxiety Rating Scale can be applied for diagnosing anxiety disorders. This scale is also useful when assessing the effectiveness of drugs taken in anxiety disorder therapy or the effectiveness of psychotherapy [23]. The Nottingham Health Profile, Short Form 36 Health Survey Questionnaire, EuroQoL questionnaire and Australian Assessment of Quality of Life are examples of general questionnaires, which have been used in clinical studies for assessing HRQoL in patients with cardiovascular diseases. Alternatively, many disease-specific questionnaires have been proposed (e.g. the MacNew Heart Disease Health-related Quality of Life for patients with cardiovascular disorders, Minnesota Living with Heart Failure Questionnaire for patients with heart failure, and St. George Respiratory Questionnaire for patients with chronic obstructive pulmonary disease). Recently, a Polish version of a disease-specific questionnaire for PAH patients known as Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR) has become available [24–26]. The CAMPHOR allows evaluation of three main dimensions, which assess symptoms, functioning, and QoL. The symptoms dimension incorporates 25 symptoms and is broken up into three subscales: energy, breathlessness, and mood. The QoL scale has 25 items, which focus on socialisation, role, acceptance, self-esteem, independence, and security. The activity scale has 15 items. Unfortunately, the use of CAMPHOR is limited by prohibitive license costs as well as the necessity of applying for permission from Galen Research on the basis of the submitted research project. Another available instrument is the Pulmonary Arterial Hypertension Symptom Interference Scale (PAHSIS) — an investigator-developed tool to measure the degree to which PAH symptoms interfere with patients' lives. The PAHSIS contains 17 PAH symptoms, which are assessed on a 0–10 scale. Patients rate how much each of the symptoms interferes with their lives and how much the symptoms interfered over the past month. Symptoms included in the PAHSIS measure include: shortness of breath (SOB) with exertion, SOB lying down, SOB at rest, awakening at night SOB, fatigue, difficulty sleeping, chest pain, abdominal swelling, swelling of ankles and feet, syncope, palpitations, dizziness, cough, nausea, loss of appetite, hoarseness, and Raynaud's phenomenon (cold, numbness of extremities). Initial psychometric studies demonstrated good validity and reliability of this tool [27].

PSYCHOLOGICAL ASPECTS OF PHYSICAL ACTIVITY IN PAH PATIENTS

For many patients with diagnosed pulmonary hypertension it is a problem to adapt to difficulties connected with symptoms and treatment of their disease. Every PAH patient experiences physical limitations to some degree, either in the form of necessity to rest more often or significant limitations in physical functioning. Limited physical capacity observed in this group of patients has an influence on many aspects of life. In a pilot study aimed at evaluating the role of exercise in PAH, Mereles et al. [28] randomly selected 30 patients for a training programme and for a control group. The intervention consisted of three weeks of training in hospital and a follow-up 12-week exercise routine at home. After 15 weeks, the exercise group improved their results in the area of physical capacity and QoL; in the control group, on the other hand, the parameters related to the above-mentioned areas either did not change or even worsened. The exercise routine was safe and well-tolerated by the group of patients with PAH [28]. In another study of 52 PAH patients, relations between symptoms of depression, physical activity, and the severity of their condition were analysed [29]. The patients were asked to fill in a self-evaluation questionnaire on depression symptoms and physical activity. The analysis proved a significant relationship between physical activity and depressive symptoms. The average score of patients under study in the Beck Depression Inventory was 11, which indicates mild and moderate symptoms; in the case of 21% of patients the score was higher than 16, which indicates moderate or severe depressive symptoms. Patients with moderate or severe depressive symptoms achieved much worse results in the field of physical activity as compared to patients who did not show any or showed minimal symptoms of depression. The most important conclusion drawn from this study is the fact that depressive symptoms are independently associated with physical activity of PAH patients, taking into consideration demographic characteristics and the evaluation of the disease severity and haemodynamic variables [29]. Since then, the results of further studies have confirmed that trained PAH patients achieved higher levels of physical activity, their level of fatigue was lower, and six-minute walk test was improved, as was their QoL as compared to the control group who had no physical training [30–32].

Interconnection between QoL, depressive disorders, and physical activity implies the necessity to prescribe to PAH patients a clinical procedure that promotes physical activity relevant to their clinical condition [29]. It cannot be expected that physical exercise will change the progression of PAH. However, improving the patient's strength and aerobic fitness by exercising certain groups of muscles will allow the patients to undertake physical activity with lower cardiovascular exertion. Admitting the fact that patients are able to

improve their well-being not only by taking medications may contribute to reducing anxiety and depression connected with the disease [6].

COGNITIVE DISORDERS

Cognitive disorders hinder the possibility for patients to return to work, function properly in a family, or enjoy life. Even minor cognitive disorders may result in problems with performing everyday activities, such as taking medications, driving a car, financial management, etc. Although the influence of PAH on cardiovascular activities has been well-documented, we still have little knowledge of the impact of PAH on cognitive functions, despite the fact that patients report problems with memory and concentration [33]. It is assumed that PAH patients may suffer from cognitive disorders similar to those observed in patients with hypoxaemia or heart failure. A group of researchers from Brigham Young University conducted a study among 46 adult PAH patients in order to assess their cognitive functions, depression, anxiety, and QoL using neuropsychological tests [33]. Cognitive disorders were detected in 58% of patients. Specifically, 56.5% of patients suffered from motor disorders, 41.3% from memory impairment, 17.4% reported bradyphrenia, 15.2% complained of slowness of executive functions, and 13.5% reported impaired concentration. 26% of patients showed moderate or severe symptoms of depression and 19.6% — moderate or severe anxiety symptoms. The mean duration of PAH (time from diagnosis) was 2.6 years, and 70% of these patients were in III or IV functional class [34]. It is possible, therefore, that at an earlier stage of PAH development, patients show less frequent or less severe cognitive disorders. No difference was recorded between patients showing cognitive disorders or without such disorders with regard to depression, anxiety, or QoL. Lower QoL was connected with impaired verbal and operational memory. No significant correlations were noticed between medical variables and the results of neuropsychological tests.

Summing up, doctors taking care of PAH patients should be aware of the negative impact of the disease on cognitive processes, which may be present in the majority of patients [35]. Memory impairment may be diagnosed early, using simple tests, such as the Mini Mental State Examination, Montreal Scale of Cognitive Functions, or Clock Drawing Test [36]. These tests may be carried out in any doctor's office without unnecessarily prolonging the appointment. Based on these test results, it is possible to estimate whether a given patient shows symptoms of cognitive disorders and how severe they are. If test results confirm the suppositions, specialist consultation is necessary, e.g. in the form of neuropsychological and neurological tests, and this fact should be taken into consideration when planning a therapy for a given patient.

FROM ADOLESCENCE TO ADULthood — TRANSITION OF A PAH PATIENT FROM PAEDIATRIC TO ADULT CARE CENTRE

A very important but frequently underestimated problem concerning the therapy of PAH patients is the moment when teenage patients are taken over by cardiologists who treat adults [37]. It is a difficult moment for medical teams, young patients, and their parents. The organisation of the healthcare system in Poland is such, that patients with "adult" conditions remain in the care of a paediatrician until the age of 18 years, when they are automatically transferred into the "adult" healthcare system. Usually, nobody is really prepared for such a transition. A change of medical centre is a systemic change: a change of the place of treatment, healthcare system, as well as methods and style of communication between a doctor and the patient [38]. In a healthcare centre for adults, a teenager is treated as an adult. Young patients are thus expected to be mature and capable of solving problems; they are required to take independent decisions, e.g. choosing a treatment method. As an adult, the patient receives less emotional and informational support from a doctor and medical staff. Although a young PAH patient usually knows a lot about his or her disease and treatment, there is a risk that, feeling very independent, they may undertake some therapy on their own or modify the prescribed pharmacotherapy. The family of a young patient also needs to adapt to the change. A problem that is specific to parents is the fact that they need to accept their supporting role in the treatment of their child. Parents often feel excluded or threatened when a doctor does not communicate with them, but focuses on the relation with a young patient. A consequence of such changes in the adaptation period may be a disturbed feeling of safety, distrust towards the healthcare unit or a particular doctor, pressure exerted in order to change therapeutic methods, or making attempts to return to a paediatric centre.

Another problem occurs when a paediatrician who has been taking care of a patient for many years finds it difficult to stop participating in the treatment process. It may result from the feeling of responsibility for a patient and from the fear that his or her therapeutic concepts will not be continued by another doctor. The physician unconsciously conveys his or her fears when talking to the patient and the patient's parents, and may express concern about what will happen to the patient when they are transferred to a healthcare unit for adults. Sometimes the doctor insists on prolonging the therapy at the healthcare unit for children and encourages the family to apply for such a prolongation [39]. The experience of medical centres in Belgium shows that after the first appointment with a cardiologist taking care of adult patients, young patients suffering from PAH and their parents had the impression that the

doctor knew nothing about their disease, because the volume of medical records which the doctor had at his or her disposal was limited when compared to documents gathered by the paediatric centre. It has been established, that an element that contributes to a successful transfer of a PAH patient to a healthcare unit for adults, is making a compilation by the patient and their family of his or her medical history. Literature suggests that special programmes should be developed to facilitate the transfer of young patients into the care of medical centres for adults. All medical staff, i.e. not only doctors, are trained in this area. It is suggested that e.g. a position of a coordinator for the patient's hand- and takeover should be established (it would be a person responsible for organising patient handover and monitoring his or her treatment after the patient is taken over by a healthcare unit for adults). Administrative and legal solutions as well as criteria for establishing the readiness of a patient to be transferred and determining the moment of takeover are being developed [38]. Transfer of PAH adolescents into the care of cardiac centres for adults is a complicated process, that requires careful preparation. PAH progress in the case of every adolescent has its own characteristics, dynamics, and development and is connected with a number of restrictions and responsibilities. It leads to certain consequences for the child's functioning: it affects the child's psychophysical condition, intellectual capabilities, emotional state, and social functioning. Therefore, each adolescent suffering from PAH should be treated individually. Each patient's therapeutic needs ought to be systematically recognised, and attempts should be made to create an environment satisfying such needs.

THE ROLE OF FAMILY IN PAH THERAPY

It is a difficult task for the medical team taking care of PAH patients to focus not only on the patient, but also on the patient's family. The family plays two important roles in the therapeutic process: motivational and functional. It motivates a patient to continue the therapy because this gives a sense to his or her life, and the functional role of the family comprises, for example, reminding the patient to take medications, to go to a follow-up appointment, etc. On the other hand, the family may of course create obstacles to effective treatment. The main problem is lack of understanding and lack of awareness of how important certain medical recommendations or restrictions connected with the disease are. Inadequate treatment not only leads to long-term and/or final consequences, which are difficult to accept, but also to short-term ones, which affect the everyday functioning of patients and their families [40].

At the same time, almost two thirds of patients with PAH report that the disease has an impact on their sexual life and libido, which is partially caused by low self-esteem, and partially by physical limitations brought by PAH. The impact on family life is evident, manifested among others by the fact that pregnancy is contraindicated for patients in PAH because the

risk is high both for the mother and the foetus [41]. In addition, more than half of the patients noticed that PAH affects their ability to play with their children or grandchildren, or the amount of physical activities undertaken in their spare time; 77% of PAH patients reported the influence of the disease on their capability to travel or go on holiday [42, 43].

When considering care of PAH patients, it should be recognised that the burden of the disease is felt not only by the patients themselves, but by their carers as well. The role of a carer is often played by the patient's spouse, partner, or other family members. As well as patients, the study carried out by the European Pulmonary Hypertension Association included also 129 carers (a carer was either a spouse/partner [55%], a parent [21%], a child [18%], a family member [5%], or a friend [1%]). The study showed that more than a half of carers (57%) reported a significant impact of the disease (8–10 score on a 10-degree scale) on their everyday life. The percentage of carers experiencing such a high level of impact on their life increased with worsening of a patient's functional class (41%, 53%, and 88% of persons taking care of patients in II, III, and IV functional class, respectively) [39]. A large number of carers (43%) reported that additional tasks and responsibilities connected with the patient's condition were strenuous for them. As many carers believe that they are not prepared to take care of patients, it seems that the first step to be taken in order to reduce the burden experienced by them in connection with the course and treatment of PAH should be to educate the carers on the patient's condition and the ensuing consequences of the disease [42].

PALLIATIVE CARE

Patients suffering from chronic diseases with poor prognosis are offered a specific, holistic management, known as palliative care. Although it is often associated with care of patients suffering from cancer, it is not a synonym of terminal care, and it should become an integral part of procedure in the case of chronic and life-limiting diseases [6] because it improves the QoL of patients and their families. Focus is placed not only on helping the patient live for as long as possible, but also on providing optimal supportive care, including prevention and reduction of pain and disability by early diagnosis, assessment and treatment of pain and other physical, psychosocial, and spiritual problems [6]. Literature on PAH does not currently include any comments on palliative care, and little attention is paid to this issue. It is suggested that such a special attention should be paid to patients who "may quite probably die" or may require repeated hospitalisations due to acute medical complications connected with the disease, so that the medical team would be more pro-active in providing information that is needed in order to take reasonable end-of-life decisions [44]. Appropriate moments for such steps should be established. It is important, however, that the medical team should ensure the opportunity to discuss such difficult issues

with a patient suffering from PAH before the patient's clinical condition deteriorates and when the patient is ready to have a rational conversation about issues connected to the end of life (functioning, complications, QoL, care needed) [6].

CONCLUSIONS

Pulmonary arterial hypertension significantly reshapes the personal and professional lives of patients due to its chronic nature, varying clinical profile, and uncertain perspectives of treatment. PAH is at this time an incurable disease, which requires consideration not only of the strictly medical dimension, but also of the psychosocial aspects of care. It is a challenge connected to insecurity and anxiety as regards the course of the disease, ensuing treatment, and prognosis. It leads to changes in almost every area of human life (physical, emotional, social, professional) and imposes new, additional responsibilities, which the patient sometimes needs to manage on his or her own. At present, psychological care of PAH patients is still under development. Co-ordinated efforts are required in the area of research, so that the QoL and the nature of emotional disorders experienced by PAH patients and their carers can be better understood [45]. Such improvement would allow us to decide whether standard treatment is effective in this group of patients and how the management of the disorders discussed above would help to prolong patients' lives and improve their QoL. Prompt diagnosis and relevant psychological treatment of emotional and cognitive disorders in PAH patients may contribute not only to improving the QoL, but also to improving the results of therapy.

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