

Common problems in rare congenital heart diseases

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Congenital heart disease (CHD) is the most common developmental anomaly and is found in about 1% of all live-born children. Recent progress in cardiac surgery and pediatric cardiology has resulted in large numbers of adult patients who have undergone surgical correction of complex congenital heart defects. It is estimated that about 85% of newborns with heart defects will reach adulthood. The 32nd Bethesda Conference October 2000 guidelines indicate that the frequency of CHD in adults is about 2800 per million subjects, and in at least half of these, the disease is moderate or complex [1,2]. In 1996, the number of adult patients in Canada with congenital heart disease was 94 000, increasing to 124 000 in 2006. In the USA, it is estimated that each year the number of adults with congenital heart disease will increase by about 9000 cases, reaching 90 000 cases within 10 years. Among adults with congenital heart disease, the number of patients not undergoing any intervention (cardiological or cardiac surgery) is decreasing, owing to the growing population of subjects submitted for interventional cardiology procedures or one- or multi-stage cardiac surgery.

The ESC guidelines recommend multispecialist care including pediatric specialists (pediatric cardiologists, pediatric cardiac surgeons) and specialists caring for adult patients (cardiologists, cardiac surgeons, anesthesiologists, internists, surgeons). Psychological support and social assistance play an important role in the management of such patients [1,3,4]. The population of adults with congenital heart disease is heterogeneous with respect to the type and complexity of the defect as well as the clinical manifestations. Many patients lead normal adult lives after having undergone corrective surgery for congenital heart disease in their childhood, however, a percentage of them will still require multispecialist care.

These adult patients with moderate and extremely complex CHD, despite reaching adulthood, are unable to function independently in society [5]. CHD affects their everyday life in many

different ways. Some patients are inclined to believe that they are “different” from the rest. Very frequently they are raised by over-protective parents. Symptoms such as cyanosis, changes in their fingernails, or scars make them perceive their own body as being far from normal and physically less attractive. Their relationships with friends are frequently dysfunctional, they frequently miss lessons, sometimes pursue individual educational plans, do not participate in elective courses, and experience reduced exercise tolerance. The feeling of being different is usually stronger during adolescence. This may lead to developmental, emotional, and social problems in some adolescents and adults with congenital heart disease. Some of these include memory disorders, attention problems, difficulty in planning, and impaired intellectual and educational development. These problems are partially related to the impact of society, thus contributing to psychological difficulties and decreasing self-confidence [6].

Previous studies have shown that adolescents and adults with congenital heart disease have higher levels of psychological distress and behavioral problems. Possible factors which are related to higher psychological distress include: cardiac status, health related quality of life (HRQoL) and self-esteem of diseases severity. Earlier studies showed that patients with congenital heart diseases have lower self-esteem. Additionally, it is known that psychological factors such as personality, behavior, emotions, and cognitive processes may affect body responsiveness and modify the course of various diseases, including cardiovascular disease [7,8,9,10]. Furthermore, the physical capacity of patients late after corrective surgery is lower than the general population, being far from meeting the criteria of full recovery. Because of this, such patients have anxiety towards performing physical activity, which can hinder participation in social life [11,12,13].

These patients often have disability certificates and are financially dependent on their parents and social welfare. Moreover, when

entering adulthood without adequate preparation, they often have trouble coping with stressful situations such as deciding to get married, finding a job, etc. Actuarial data show that the level of professional activity among young people with disabilities in Poland is alarmingly low. Only one out of five of people with a disability up to 24 years of age is employed or seeks employment, while as many as one third of young disabled people exclude themselves from working after completing their education. Seventy two percentage of young people with disabilities did not earn their own money, 80% described themselves as retired, and 40% chose to keep their disability pension instead of taking up a job. Being employed carries with it the risk of losing one's social pension. This is seen as an enormous obstacle when trying to motivate people in this patient group to start a new job. Being employed carries with it the risk of losing one's social pension. This is seen as an enormous obstacle when trying to motivate people in this patient group to start a new job. Moreover, there is often a lack of information on available employment opportunities for this patient group. Additionally, vocational education programs frequently do not meet local labor needs and do not take into account the type of disability in these patients [3]. Extended periods of unemployment can have a damaging effect on young people. They lose contact with friends, become socially isolated, lack self-confidence, have low incomes, experience stress within the family, intellectual and spiritual capacity deteriorates, have physical and mental health disorders, have limited access to the housing market and methods of transportation, have insufficient professional mobility, and difficulty in finding a new job. Furthermore, those who are unemployed often have a less healthy diet than others, lose interest in acquiring knowledge, and are frequently prone to developing dependencies towards addictive substances. Inadequate financial resources and increasing health-related expenditures cause greater psychological distress. Health and life insurance may also create additional problems [14,15,16]. Professional inactivity in such a large group of young people may also impose a burden on the economy and society. National expenditure on pensions (disability, social, rehabilitation, and disease) and costs of disease-related inability to work frequently exceed direct costs related to diagnosis and treatment of the defect. These costs are usually covered by employers, which can also be a hurdle to employment of young people with disabilities.

Tremendous progress in medicine, especially pediatric cardiac surgery and cardiology, has resulted in large numbers of young adult patients who have complex congenital heart defects corrected through surgery [17,18,19,20]. This patient group is steadily increasing all over the world and will require a different management strategy. In addition to medical care provided in specialized centers, it is essential to establish a model of professional mobilization for these young people, to identify individual needs, and to provide motivational and psychological support for those returning to work. Increased occupational activity of young adults with congenital heart disease and improved core competences, interpersonal communication skills, and self-presentation as well as acquiring coping skills will improve the life quality and physical capacity in this patient group. Taken together, these measures may improve the prognosis of these patients.

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