

Adolescents with congenital heart diseases

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Abstract

The remarkable improvement in pediatric cardiosurgery observed over the last few decades has meant that many patients with congenital heart diseases have entered adult life. The growing population of adults with congenital heart defects (ACHD) requires specialized treatment. The team of specialists should consist of cardiologists experienced in congenital heart diseases, cardiothoracic surgeons, obstetricians and anesthesiologists. The need for providing ACHD with appropriate healthcare remains largely unmet. The proper care of ACHD requires an excellent knowledge of defect residuals and post-operative sequel complications. The commonest clinical problems are arrhythmias, impulse formation and conduction disorders as well as heart failure with complicated pathophysiology. The medical management of these patients is mainly based on doctors' experience, since Evidence Based Medicine recommendations do not cover the above mentioned clinical issues. Cyanosis must be distinguished as another clinical problem for which treatment requires special knowledge. For many women with congenital heart diseases, pregnancy and delivery may be hazardous. To date, observations show that most patients with congenital heart diseases enjoy a full life. (Cardiol J 2010; 17, 1: 11–19)

Key words: arrhythmia, impulse formation and conduction disorders, cyanosis, heart failure, pregnancy

The growing population

The extensive progress in cardiology and pediatric cardiac surgery seen in recent decades has caused more than 85% of children born with congenital heart defects (CHD) to reach adulthood. In order to provide such patients with specialized health care, attempts are being made to estimate the discussed population. Congenital heart defects occur in 4 to 10 per 1,000 live births [1]. According to Jane Somerville [2] 50% of patients with complex defects, and 90% with simple ones, reach the eighteenth year of age. Based on this assumption, estimates of the number of adults with CHD have been carried out in certain countries [3, 4]. Additionally, knowing the number of patients under care, it has been proved that specialized treatment for these patients, even in highly developed countries,

is definitely insufficient. In most countries, care does not exceed 50% of needs [1–3]. In western Poland, for instance, the estimated covered needs reached merely 10% [4]. Marelli et al. [5] were the first to provide actual numbers, rather than estimates, thanks to data from the province of Quebec covering the years 1985–2000. They have drawn several conclusions based on these numbers: 85% of children with CHD reach maturity, adults with heart defects outnumber infants with similar anomalies, and the age of the analyzed population increases systematically, with most of the patients reaching the fourth decade of life. About 5% of patients reach 60 years of age, which inevitably leads to the occurrence of additional age-related illnesses [3, 6].

The researchers from Montreal [7] analyzed the demand for medical care among adults with CHD and came up with the following findings: 87%

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of patients visited outpatient clinics, 51% required hospitalization with some of them necessitating intensive care (16%). The study also revealed that patients before 65 years of age required specialized treatment considerably more often than the remaining population of cardiac patients, which emphasizes the need for comprehensive medical care [8]. It was also estimated that a cardiologic center dedicated to adults with CHD should meet the demands of a 4–6 million population. The staff in such centers must constitute cardiologists who are experienced in the discussed issue, as well as cardiac surgeons and interventional cardiologists who have knowledge of heart defects anatomy, expected complications, and post-operative residuals, all of which enables the performance of necessary surgery. It is also essential for the treating team to include obstetricians and anesthesiologists familiar with CHD, as well as counseling psychologists and a well-trained nursing team [9].

What increases the complexity of the problem is the variety of anatomical heart anomalies present in the adult population with congenital heart disease. The most numerous group consists of patients who were operated on in the past. Within this group, there is a further distinction between those treated with older or newer surgical techniques, which bring about different clinical outcomes. The results of the primary and contemporary ways of defects correction are indeed incomparable [10, 11]. Another group consists of patients who were not subjected to cardiac surgery because it was unnecessary. Another group comprises patients where there was no possibility of surgery, or there were contraindications to surgery, of which pulmonary hypertension was the most frequent [3, 6, 9, 12]. At present, primary surgeries (e.g. Fallot tetralogy repair) are performed in specialized centers. However patients are more and more often undergoing re-operation. This may be a scheduled surgery being the next stage of cardiac surgery (e.g. Fontan operation after Glenn procedure), unavoidable like a consecutive surgery in ever-progressing disease (e.g. implantation of aortic prosthesis after original valvulotomy), or unexpected operation (e.g. renewed septal defect, pulmonary stenosis or aortic regurgitation after arterial switch operation) [13].

Cardiac surgery performed in adults with CHD requires of an operating surgeon special skills, excellent knowledge of a defect's anatomy, way of performing the surgery, and the type of possible complications from the previous surgery. Overall perioperative mortality ranges from 2% to 5% and increases with each subsequent surgery as some

patients undergo repeat sternotomy [12]. The Canadian authors proved what could have been intuitively predicted: that significantly better post-operative results are achieved when operations are performed by cardiac surgeons acquainted with children's cardiac surgery [14].

Clinical complications — residuals and sequels

Rational care for adults with CHD may turn out to be demanding as it requires not only exact knowledge of a defect's anatomy and means of correction, but also the ability to predict its hemodynamic consequences appearing within a specific period of time. Such care demands familiarity with potential post-operative complications and anatomical remnants of defect, so called *residua* and *sequelae* [2, 15–17].

Arrhythmia

The commonest clinical problems, and so the commonest cause of death in the discussed population, are heart rhythm disorders. Usually they are the first sign of hemodynamic abnormality. They occur in up to 40% of patients after 50 years of age, which in the light of an aging population creates an essential therapy problem. Arrhythmia, for instance, judged to be mild in the so called average population, may cause substantial clinical consequences in adults with CHD. This is why understanding its pathophysiology and ability to treat it are so crucial [3, 15, 17].

Supraventricular arrhythmia has been viewed as the 'price' of long-term survival after surgery performed in the past. Modern operations decrease the incidence of these complications [17]. Examples are: replacement of Sennig/Mustard operation with an arterial switch method [18]; modifications of Fontan procedure such as total cavopulmonary connection which do not remodel right atrium; and avoidance of transannular patch in the right ventricle outflow tract in tetralogy of Fallot that reduces arrhythmogenic pulmonary regurgitation [19, 20]. In general, pathogenesis of supraventricular arrhythmia in adults with congenital heart defects is compound and stems from a) primary alterations in conductive fibres route; b) enlargement of atria causing dilation, fibrosis, increased wall stress and finally inhomogeneous electrical activity; c) presence of surgical scars that favor delay in impulse conduction resulting in intra-atrial re-entrant tachycardia (IART); d) deterioration in ventricle

function: supraventricular arrhythmia is the effect of abnormal hemodynamics and can also lead to further exacerbation in heart failure; and e) sinus node dysfunction which promotes re-entrant arrhythmias [16, 17, 20–22].

The peculiarity of anatomical anomalies causes different incidence of certain supraventricular arrhythmias, in particular congenital heart defects. For example, arrhythmia resulting from accessory pathways is typical of Ebstein's anomaly and congenitally corrected transposition of great arteries. Intra-atrial re-entrant tachycardia, arrhythmia clinically resembling slower atrial flutter, occurs usually in patients with both transposition of great arteries after Mustard/Sennig operation and/or univentricular heart after Fontan procedure. The least frequent in this group of patients is atrial fibrillation, which is typical of 'left' heart diseases, such as defects of aortic or mitral valves [17, 19, 23, 24].

As it follows from the pathogenesis presented above, treatment of supraventricular arrhythmia should in the first place entail correction of possible hemodynamic complications. In the case of a sudden onset of atrial fibrillation or flutter, cardioversion is the most recommended initial treatment of choice despite high risk resulting from incomplete anticoagulation therapy [2, 17]. Pharmacotherapy in patients with CHD is mostly extrapolated from average population since there are no available studies based on Evidence Based Medicine (EBM) recommendations [17, 25]. It is important to avoid negative inotropic agents: thus digitalis and amiodarone would be recommended. On the one hand, those drugs should be administered with caution, especially in cyanotic patients; but on the other hand, amiodarone is known to be the most successful agent in preventing supraventricular arrhythmia recurrence, so dangerous in the discussed population [3]. In spite of full pharmacological treatment, however, clinical practice shows that IART recurs in 70% of patients after two years [26]. Implantation of a pacemaker is yet another preventive measure undertaken against supraventricular arrhythmia, as the device eliminates excessive bradycardia and prevents its intensification during pharmacotherapy [27]. Catheter ablation ranks as a modern and widely used form of treatment in the above mentioned arrhythmia. It must be emphasized that this procedure should be carried out exclusively in highly-specialized centers because it requires exact knowledge of both a defect's anatomy and its former surgical repair.

Unfortunately, therapeutic effects are still not satisfactory: IART ablation success is almost 90% but this is a short-term outcome. Also, primarily successful ablation of accessory pathways in Ebstein's anomaly ends up with arrhythmia recurrence in 25% of patients within months. Ablation is least effective in patients with univentricular heart after Fontan procedure [26–28], most likely due to anatomical complexity of the defect. In treatment of supraventricular arrhythmia, consideration should also be given to surgical ablation during atrial maze operation. However, any decision about surgery must be well thought out as it entails undergoing another operation and the recurrence of arrhythmia still scores several per cent [17, 19].

As for the occurrence of ventricular arrhythmias, they are rare among adults with congenital heart defects, and mostly recorded in patients after Fallot repair and with left ventricular outflow tract defects [17, 24]. Such arrhythmia may contribute to sudden cardiac death [15, 16], which affects 5% of patients after Fallot repair [29]. The pathogenesis of ventricular arrhythmia in patients with congenital heart defect is still under investigation. However, it is assumed that ventricular arrhythmia results from: a) presence of scar after ventriculotomy creating substrate for re-entry circuit; b) fibrosis of myocardium due to long lasting cyanosis; c) valvular regurgitations causing ventricle's dilation, increased stress of its walls and inhomogeneous electrical activity regions; and d) way of myocardium protection during operation. Moreover, in patients with Fallot's tetralogy, a contributory factor may be pulmonary regurgitation and right bundle branch block that conduce re-entry circuit, especially in the presence of a scar [17, 30]. Treatment of ventricular arrhythmia, similarly to supraventricular, requires, in the first place, hemodynamical correction of the defect (e.g. valve replacement, closure of residual shunt or widening of conduit) [3, 15, 19, 20]. Pharmacotherapy is applied according to the same principles as in supraventricular arrhythmia treatment, where experience is extrapolated from the average population and treatment is based on the exclusion of negative inotropic agents. There is still no appropriate data resulting from studies based on EBM available. Ablation procedure is performed in rare cases (in tetralogy of Fallot short-term success rates are 80%, with recurrence amounting only to 20%) [26, 31]. There is increasing use of implantable cardioverter-defibrillators (ICD). Such devices are applied predominantly in patients after Fallot repair, with congenitally cor-

rected transposition of great arteries and complex aortic defects. The young age of patients and frequent sinus tachycardia, however, are likely to result in the numerous inappropriate shocks that were observed in 25%. In adults with CHD, ICD has proven value in secondary prevention. Nonetheless, clinical features justifying primary prevention are still sought in this group of patients [32].

Impulse formation and conduction disorders

A serious and frequent problem pertaining to the analyzed population is usually impulse formation and conduction disorders. Congenital anomaly of sinus node is observed in heterotaxy syndrome, whereas acquired type of this dysfunction is characteristic of both Mustard/Sennig operation for transposition of great arteries and Fontan operation [17]. Congenital atrio-ventricular block develops most notably in common atrio-ventricular canal, congenitally corrected transposition of great arteries, and in Ebstein's anomaly. Acquired atrio-ventricular block may result from closure of ventricular septal defects, surgery for left-heart outflow tract obstruction, as well as aortic valve replacement [16, 33]. Some of these patients require permanent pacemaker implantation.

This surgery, similarly to other procedures mentioned above, calls for special skills on the part of the cardiologist. Performing the procedure, electrophysiologists must take potential obstacles into consideration: a) persistent left superior vena cava and often concomitant extreme dilation of coronary sinus changing the route of lead implantation; b) the possibility of post-operative narrowing of superior vena cava; and c) absence of venous chamber access in patients after Fontan repair. Because of those hazards, an epicardial approach is still applied in many patients. This procedure, nonetheless, is not free of risks: greater probability of lead damage, higher stimulus threshold which is decreased through the application of steroid-eluting features, and finally the necessity of thoracotomy that is risky, especially in cyanotic patients [34]. Due to its physiological pacing mode, particularly dual-chamber, the endocardial approach would be preferable [33]. Implanting ventricular leads is yet another challenge, especially when treating patients after Mustard/Sennig operation. They may sometimes need dilation of tight obstruction along the upper limb of the baffle. It is also vital to place the lead properly in morphological left ventricle. An effective solution for difficult anatomical anomalies

seems to be an appliance of hybrid stimulation, where one lead is epicardial whereas the other is placed into the coronary sinus [26]. Furthermore, it must be borne in mind that patients with intracardiac shunts who require endocardial pacing must undergo anticoagulant therapy [35].

Heart failure

The most frequently observed symptom in adults with CHD is heart failure. Defining heart failure in this group of patients is particularly difficult. The state may be caused by deterioration in function of only right or single ventricle, shunting of arterial and venous blood, and/or increased pressure and resistance in pulmonary circulation. All these clinical features decrease exercise capacity and influence outcomes of objective oxygen consumption parameters [36]. Patients with CHD easily adapt to exercise intolerance, and by reducing daily activities they miss symptoms of successive deterioration in physical efficiency. This improves their daily quality of life, but on the other hand misleads the doctor in charge. However, such asymptomatic patients have been proved to have decreased oxygen consumption [37, 38]. Furthermore, a spiroergometric test, an objective tool, reveals lower oxygen consumption in patients with complex heart defects, cyanosis, pulmonary hypertension or atrial flutter/fibrillation [39, 40]. Pathogenesis of heart failure in adults with CHD is complex and consists of: a) decrease in contractility of heart ventricle or both ventricles; b) pressure and/or volume overload; c) myocardial ischemia resulting from coronary arteries anomalies or damage during surgical procedure like arterial switch in transposition of great arteries or tetralogy of Fallot operation; d) insufficient protection of myocardium during cardiosurgery; and e) function deterioration of morphologic right ventricle in systemic position after Mustard/Sennig operation or in congenitally corrected transposition of great arteries [36].

Pharmacological treatment of heart failure in the discussed population is derived from the scant studies that are mostly unrelated to EBM recommendations. Only the research concerning losartan [41] and carvedilol [42] meets these requirements. Efficacy of these drugs, however, was not confirmed in patients with CHD. This is why the therapy is based on generally accepted principles that are adopted in the treatment of left ventricle heart failure. However, some distinctions and restrictions must be underlined: administration of angiotensin-converting enzyme inhibitor in cyanotic patients

due to Eisenmenger syndrome may result in worsening right-to-left shunting due to concurrent reduction in systemic vascular resistance and therefore blood oxygenation decreases. Furthermore, beta-blockers alone, or in combination with an angiotensin-converting enzyme inhibitor, should be very carefully prescribed in adults with CHD, especially patients with left ventricle dysfunction, as observed in aortic coarctation, Marfan syndrome or subaortic stenosis. Furthermore, the efficacy of discussed drugs has not been demonstrated in patients with systemic morphologic right ventricle and single ventricle [43].

An isolated record shows positive results of aldosterone antagonists applied for patients after Fontan repair with protein-losing enteropathy [44]. Particular caution is required during the administration of diuretics. This mainly concerns patients dependent on preload such as adults after Fontan or Mustard/Sennig procedure or cyanotic patients where application of diuretics may lead to a dangerous increase in blood viscosity [16]. Besides, attempts at resynchronization therapy are being made. They predominantly affect patients after operation for tetralogy of Fallot and congenitally corrected transposition of great arteries. The results are promising, as improvement of heart function was observed irrespective of a left or right ventricle functioning as the systemic pumping chamber [45].

Invasive treatment is applied more and more often in adults with CHD. The benefit of that kind of treatment is prevention of thoracotomy and burden of extracorporeal circulation. Long-term observation lays basis for the claim that interventional treatment is a procedure of choice in pulmonary stenosis, ostium secundum atrial septal defect, re-coarctation of aorta and patent ductus arteriosus [46]. Treatment of native coarctation of aorta shows positive effects and its results are comparable to a surgical approach [47]. Transcatheter closure of ventricular septal defects poses a 5% risk of late complete atrio-ventricular block. That is why closure of only muscular septal defects is safe [11]. Aortic balloon valvuloplasty in the discussed population is considered only in particular circumstances, namely in case of contraindicated surgery or pregnancy. These limitations result from poor outcomes of the procedure: i.e. restenosis observed in up to 40–50% within five to ten years, and/or exacerbation of aortic regurgitation. When performed by an experienced cardiologist, good results are obtained in angioplasty of pulmonary arteries stenosis due to residuals after performed operation or palliative procedure. An alternative to surgery of an

increased risk is the possibility of percutaneous dilatation of both conduits (connecting subpulmonary ventricle to pulmonary artery after Fontan procedure) and baffle obstruction after Mustard/Sennig operation [46].

The ultimate therapeutic option is heart, lung, or heart-lung transplantation. About 10–20% of patients with congenital heart defects, at a certain stage of life, are estimated to be selected for such treatment. Survival rates, in specialized centers, are comparable to the outcomes of this therapy in the average population: they equal about 75% after one year and 60% after five years. It must be stressed that frequently distorted vascular anatomy, as observed in transposition of great arteries or after Fontan repair, creates additional intraoperative surgical obstacles [48].

Cyanosis

A particular condition in patients with CHD constitutes cyanosis. It occurs due to blood desaturation resulting from reduction in pulmonary blood flow (in primary cyanotic defects like tetralogy of Fallot or transposition of great arteries) or as the consequence of secondary pulmonary hypertension, present mostly in simple shunt lesions. Adults with primary cyanotic defects are extremely rare, as most of them undergo surgery in early childhood. The few that reach adulthood without such an operation are those in whom the balance between size of ventricular septal defect and pulmonary stenosis is preserved. The balance provides adequate tissue oxygenation and cardiac output, yet does not affect pulmonary hypertension. Secondary pulmonary hypertension is observed in 5–10% of congenital heart defects; among them the percentage of patients with complex defects after palliative procedures is still increasing [1–4, 16]. Clinical presentation of cyanosis affects multiple organs and results from secondary erythrocytosis due to the raised production of erythropoietin. Augmented haemoglobin production is an adaptive mechanism guaranteeing adequate tissue oxygenation. Increased hematocrit leads to blood hyperviscosity followed by specific clinical symptoms, such as headaches, vertigo, visual disturbances, paresthesias and myalgias.

An additional cause of some of these symptoms is iron deficiency, observed mainly in patients who underwent phlebotomy. Due to a lowered count and dysfunction of platelets, as well as disruption of intrinsic pathway (reduction of activation of II, VII, IX, X, V and von Willebrand factors), cyanotic

patients often exhibit bleeding and thromboembolic complications such as stroke, pulmonary bleeding and large pulmonary vessel thrombosis. Also, the risk of intraoperative bleeding rises substantially in such patients owing to the excessive net of small vessels. Moreover, an increased heme breakdown facilitates hyperuricemia and eventually joint and kidney changes related to gout. Impairment of hepatic function, associated with bilirubin elevation, leads to cholecystitis, often followed by cholangitis. Kidney dysfunction is often observed as a result of secondary glomerulopathy. Finally, cyanotic patients are at risk of developing infective endocarditis, cerebral abscess and pneumonia [16].

The basic procedure that guarantees rational care of cyanotic patients with CHD is a referral to a specialized center, since it is commonly known that the most effective treatment couples with experience in a given area [1–4]. The situation is that general practitioners or even cardiologists who are not trained in congenital heart defects, may encounter only a few patients with secondary pulmonary hypertension in their careers due to the rarity of Eisenmenger syndrome. As Somerville [2], a pioneer practitioner and a source of knowledge concerning adolescents with congenital heart defects aptly noted, doctors commit a gross mistake when they avoid referring discussed patients to specialists and keeping them as ‘rare stamps in an album’.

General recommendations, so essential in this group, include instructing patients on safe lifestyles which entails abstaining from physical exertion that could lower systemic pressure and enhance right to left shunt followed by decreased saturation. Besides, high pulmonary resistance hinders additional left atrial and ventricular inflow, which results in a drop in ejection fraction. Furthermore, staying in a high temperature should be avoided as dehydration increases hematocrit (and consequently blood) viscosity. This issue is worth discussing with, mostly young, patients with CHD since deaths at the disco are quite often observed in adolescents with CHD [49]. Hypovolemia resulting from diarrhea, vomiting or fever requires immediate infusion with fluids [16].

The method too often applied by general practitioners in a case of cyanosis is phlebotomy. It needs to be emphasized that this procedure is only recommended in coexistent symptoms of excessive blood viscosity observed with hematocrit exceeding 65%. This is due to its serious side effect which is iron deficiency anemia resulting in microcytosis aggravating thrombotic complications. The condition remains frequently undiagnosed as the hemo-

globin equaling 15 g/dL, in healthy patients assessed as a normal value, is however in cyanotic patients insufficient, as the border line for them approximates 18 g/dL. Contrary to previous suspicions, increased hematocrit does not lead to dangerous strokes. The risk factors for named complications are iron deficiency, arterial hypertension and atrial fibrillation [50]. During the procedure, blood withdrawal should not exceed 250 to 500 mL and additionally it should be replaced with concomitant intravenous saline volume ranging from 750 to 1,000 mL. Moreover, no more than four phlebotomies can be performed yearly. If clinical improvement does not occur, the reason may be iron deficiency anemia that necessitates the prescription of oral ferrous sulfate (325 mg daily). It is justified to perform phlebotomy preoperatively in order to improve hemostasis [16].

The issue of anticoagulation therapy remains unresolved. Due to increased thrombotic and bleeding risk in cyanotic patients, treatment with coumarin derivatives is accepted only in atrial fibrillation, intracardiac mechanical prostheses and conduits accompanied by advanced heart failure. Indications for such treatment are episodes of massive pulmonary thrombosis, observed in about 30% of patients with Eisenmenger syndrome. However, a great expert on that topic, Perloff et al. [51], is against administration of coumarin derivatives even in such settings. He claims that antithrombotic agents may lead to almost intractable pulmonary hemorrhage [52]. A frequent and life-threatening clinical complication is hemoptysis. The condition requires adequate therapeutic approach including hospitalization, physical activity limitation and cough reflex elimination. In an advanced case, fresh frozen plasma, platelet mass, vitamin K, factor VIII, or ultimately cryoprecipitate should be secured. Bronchoscopy is definitely contraindicated as it does not provide substantial data and at the same time may be a potential source of pulmonary hemorrhage. High risk of serious pulmonary infections necessitates flu and anti-pneumococci vaccination [53]. Oxygen administration does not usually lead to increased saturation but can result in dangerous airway mucosal dehydration [54].

The importance of highly-specialized centers must be also mentioned when talking about the treatment of cyanotic patients. Non-cardiac surgeries in cyanotic adults with CHD must only be performed in such centers, providing adequate anesthesia, as they account for the most frequent cause of death in this population. Anesthetics which reduce blood pressure to the least extent should be administered so as not to aggravate right-to-left

shunting and in consequence desaturation. On the other hand, a sudden rise in resistance may lead to right ventricular failure. Other dangers concern intraoperative arrhythmias, significant blood loss and bleeding complications typical of cyanosis. For that reason, local anesthesia is the preferable option. Epidural anesthesia should not supplant general anesthesia due to the increased hypotensive effect and bleeding risk. Lastly, an important role in cyanosis treatment is played by intraoperative monitoring, as it helps to evaluate volemia and pressure changes as well as pulse oximetry in order to assess oxygen saturation [55].

Pharmacotherapy of pulmonary hypertension is still at the stage of clinical analysis. Progress in the pathophysiology study of pulmonary hypertension, observed in recent decades, has resulted in the introduction of new groups of agents: prostacycline derivatives (Epoprostenol, Treprostinil, Beraprost, Illoprost), phosphodiesterase inhibitors (Sildenafil, Tadalafil), endothelin receptor antagonists (Bosentan, Sitaxantan, Ambrisentan) or nitric oxide. These drugs should be exclusively administered to patients in NYHA functional class III and IV [53]. The named drugs reduce not only pulmonary but also systemic pressure; hence they potentially increase right-to-left shunting and in turn produce diverse results in patients with Eisenmenger syndrome. For the time being, only Bosentan has proved effective in this population [56].

Pregnancy

Pregnancy and the moment of delivery pose an enormous risk for female cyanotic patients. The risk increases with the degree of desaturation. In patients with blood saturation below 85% maternal mortality reaches 50%, which is similar to the percentage of spontaneous abortion [57]. Conception is contraindicated in advanced cyanosis, which additionally constitutes one of the few cardiologic indications to pregnancy termination [58]. Such a dramatic decision is also justified in patients with advanced heart failure, severe aortic stenosis, and in Marfan syndrome when aortic root diameter exceeds 45 mm [59]. Pregnancy in patients with other congenital heart defects is usually well tolerated, although it may become problematic due to hemodynamic changes. During pregnancy, increased blood volume and tachycardia, as well as decreased systemic vascular resistance, are observed [60]. Metaanalysis reported on about 2,500 patients with congenital heart defects [61] showed that serious cardiologic complications, like significant arrhyth-

mia, heart failure, myocardial infarction, stroke, or even death, were noticed in 11% of women, mostly patients with the following conditions: cyanosis, univentricular heart, congenitally corrected transposition of great arteries after Mustard/Sennig procedure, truncus arteriosus, common atrio-ventricular canal and after Fontan repair. Spontaneous abortions occur more frequently than in the average population (15% vs 10–12%). Therefore, a multidisciplinary approach including the assistance of experienced cardiologists, obstetricians and anesthesiologists is required [62]. The risk of possible complications during pregnancy and delivery is reduced by adequate cardiologic assessment and possible correction of existing hemodynamic disorders before conception. It is also crucial to discuss with young women possible complications and the probability of heart anomaly inheritance [63]. Pregnancy should be strongly discouraged particularly in the above mentioned cases; however no available contraceptive method is optimal in this setting. The adverse effects of oral contraceptives should be borne in mind as an estrogen-related raised risk of thrombotic complications and a progesterone-related increase in blood volume may exacerbate symptoms of heart failure. However, contraception in high-risk patients is less risky than possible pregnancy [64].

Survival

The degree of hazard in a given population is measured by the number of recorded deaths. The discussed population constitutes mainly adolescents; thus mortality rate in this group is lower than in other groups with frequently analyzed cardiologic diseases. The data shows mortality in patients with CHD is relatively low and averages about 4.5% after tenyears of follow-up; only in the group of complex defects does it exceed 10% [65]. Sudden deaths have been proved to occur 25 to 100 times more often than in the average population. Moreover, this negative outcome may even appear in the second decade of life, mainly in patients with cyanosis and after systemic ventricle outflow repair [66]. The exact data collection and analysis concerning survival rate in patients with congenital heart defects are cumbersome, due to the small number, great variety of population, as well as frequent out-of-hospital deaths. Attempts are being made to determine the factors of death risk in this population. Some researchers suggest age and cyanosis [67] as the important factors, others point to post-operative complications or declining heart failure [49].

Challenges

Regardless of all the symptoms discussed above, due to the earlier efforts of pediatric cardio-surgeons and pediatricians, the great majority of adults with congenital heart defects experience a full life. This is why close and careful medical management, as well as collection and study of data, are so crucial.

Rational care for adults with congenital heart defects constitutes a great contemporary challenge, similar to the one posed by creators of new cardio-surgical techniques, as to how best to prolong the effect of their brilliant work.

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