Clinical Psychology and Congenital Heart Disease

Lifelong Psychological Aspects and Interventions

Edward Callus
Emilia Quadri
Editors

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Forewords by
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Foreword

The concept of health care as pure therapy has changed due to the discoveries in the last 20 years. The overall goal of medicine has shifted towards an approach that is concentrated on health, meant as a necessity requiring the availability of a service which is more focused on the patients as persons and on their multiplicity of needs. Medicine cannot be considered only in terms of "survival" anymore, but in a more broad perspective of the meaning of existence, which also encompasses and enhances the subjective psychological aspects of health itself.

A development of the process which involves the humanization of health care structures can be seen in the national and international hospital organization, and this can also be observed from the improved qualification and awareness of the medical and nursing staff.

It is therefore important to give the necessary relevance to the humanization of health care, where there is not only the cure of the pathology, but also attention to the global, psychophysical well-being of individuals.

The primary objective of the San Donato Hospital Group is for patients to be considered at the center of health care, being respected in all their dimensions including their personal stories, their culture, their emotions and feelings.

The IRCCS (Istituto di Ricovero e di Cura a Carattere Scientifico – Scientific Institute for Research, Hospitalisation and Health Care) Policlinico San Donato University Hospital is one of the three IRCCS of the San Donato Hospital Group: the Group is constituted by a total of 18 hospitals, and it is currently one of the first hospital groups in Europe, besides being the most important one in Italy.

Apart from being the most important reference point in Italy when it comes to cardiac pathologies, it is one of the outstanding centers in the world for the treatment of pediatric and adult congenital heart disease. In fact, in 2013 alone, approximately 550 cardiac catheterisms (both interventional and diagnostic) and 460 heart surgeries were performed for the benefit of patients affected by this condition.
It is a source of great pride for me to present this volume, the first of its kind, designed and developed at our Policlinico San Donato.

San Donato Milanese, Milan, Italy

Gabriele Pelissero
President of the San Donato Hospital Group

Foreword

For several years in our clinical practice, we have observed the psychological support to patients and families in the Pediatric and Adult with Congenital Heart Disease Center of the Cardiovascular Center "E. Malan" of the IRCCS (Istituto di Ricovero e di Cura a Carattere Scientifico – Scientific Institute for Research, Hospitalization and Health Care) Policlinico San Donato University Hospital. For the patients and their families who are involved in the difficult path of dealing with complex pathologies, like some conditions of congenital heart disease can be, it is essential to be adequately welcomed and listened to.

When it comes to providing assistance to hospitalized children, the necessity for humanization manifests itself more strongly. In fact, the hospitalization process can have important repercussions on the psycho-developmental processes in the child.

This happens because children and their families find themselves having to handle the condition and all that its treatment involves, which is often inevitably traumatic. Furthermore, they are often distanced from an environment which they are familiar with. There are also occasions in which children or adolescents are also necessarily separated from one or both parents for some time during their hospitalization. Having a congenital heart disease can also entail dealing with lifelong health issues during adulthood, which can have an important impact on all areas of life.

The objectives of the humanization of the hospital setting and its integration with the surrounding territory have led us to conduct a series of psychological and cultural projects coordinated by two psychologists and psychotherapists (Dr. Emilia Quadri and Dr. Edward Callus), which also include research activities, in collaboration with the NGO AICCA (Associazione dei Cardiopatici Congeniti Adulti – Italian Grown Up Congenital Heart Disease Organization).

That is why I am very pleased to write the foreword for this book, in which the psychological aspects linked with congenital heart disease in all stages of life are considered.

San Donato Milanese, Milan, Italy

Mario Carminati
Director, Pediatric and Adult
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Preface

Although congenital heart disease is the most common inborn defect, with an approximate prevalence of eight newborns for every 1,000 births, it is a not well known condition in the general population. One possible reason for this is that in the past these patients used to carry a very poor prognosis; however nowadays most of them survive reaching adulthood.

In fact, the number of adults with congenital heart disease is growing rapidly due to the advances and triumphs of cardiovascular medicine and surgery in the twentieth century. Since this population is on the constant increase, it is especially important to concentrate research efforts on the improvement of the management of these patients, including psychological aspects as well, which are an essential component of care in medical and chronic conditions.

Like all the other pathologies, although technological, scientific and medical progress has brought many benefits, it also entails the risk of potentially developing less personal and less humanized processes, systems and institutions, especially when it comes to medical institutions. Specialized psychological care in institutions, in which these patients are taken care of, is not to be taken for granted – there is a huge diversity of situations in Europe and the world.

In our role as psychologists and psychotherapists at the IRCCS Policlinico San Donato University Hospital with patients with congenital heart disease of all ages and their families, we have often asked ourselves in which way the hospitalization experience could be improved. In this context there is often a contact with suffering and emotions, which are often labelled as "negative".

In these situations, some of the conditions which cause immediate suffering cannot be changed, but a lot can be done when it comes to the attitude towards suffering. One of our main objectives was to create an environment where it was possible to legitimate and give the possibility to express what one was going through.

This was possible also through the collaboration with the nonprofit organizations in this sector, both locally and on a European level. Also in this case, our contribution was that to help create occasions, also outside of the hospital setting, in which problems can be shared, and support given, focusing not only on what is wrong but also on the incredible resources and potential of these patients and their families.

This book was conceived as a guide for the psychologist and other professionals who deal with these patients and their families. Firstly, the medical condition of congenital heart disease is described and the humanization of the health care context in general is explored. In the following parts of the book, the psychological characteristics of patients with congenital heart diseases are considered from childhood to adult age, focusing in particular on specific issues, which emerge as important in this population; for example, neuropsychological aspects, psychological functioning, quality of life, life experiences and end of life care. Apart from citing the most important literature on the various topics, there has often been an effort to describe clinical case studies, in order to give specific indications when it comes to the management of these patients.

There is plenty of literature regarding the psychological aspects of acquired heart disease, also under the name of psychocardiology. In this textbook the reader will find the results of an effort to find which aspects of psychocardiology might also be applied to congenital heart disease patients. In addition, specific examples of psychological interventions are described, such as Medical Art Therapy for these patients. Finally, the importance of associations and peer to peer support is outlined, by focusing on the development of European associations and by exploring specific examples of peer to peer support.

We are very happy and thankful that some of the top experts in the psychosocial management of these patients have contributed to the creation of this manual, and we sincerely hope that it will be a valuable tool, which improves the care provided to these patients.

San Donato Milanese, Italy

Edward Callus
Emilia Quadri

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Part I

Congenital Heart Disease and Humanization of Healthcare

Congenital Heart Disease: A Medical Overview

1

Massimo Chessa and Francesca Romana Plucchinotta

Congenital heart disease with an approximate incidence of 1 % and a prevalence at birth of 5–11 per 1,000 live births [1] is the most common inborn defect. By definition, "congenital" heart disease means usually a disease that has been present since birth. However, a wide variety of defects can be either not present or not evident from birth. Those present but not usually detected in early life include lesions such as a moderate size atrial septal defect. Others that are only anatomically present in later years with a latent predisposition prior to this, such as subaortic stenosis or many of the cardiomyopathies, are not strictly "congenital" but are often included in this group.

The most important physiopathologic aspects of the congenital cardiac malformations are the presence of shunt between arterial and venous blood and presence or absence of cyanosis.

A shunt occurs when there is abnormal communications between two chambers or two vessels. It may be described as right-to-left, left-to-right, or bidirectional. The direction of the shunt is controlled by the pressure differential between the two cardiac chambers. Most commonly the blood goes from the high-pressure left side of the heart to the low-pressure right side of the heart resulting in a left-to-right shunt. The consequences of this shunt are (1) excessive blood flow into the lungs causing shortness of breath and increased pulmonary vascularity on a chest x-ray, (2) increased volume overload of one ventricle resulting in hypertrophy of myocardium and chamber dilatation, and (3) turbulence of abnormal blood flow producing a heart murmur.

Untreated left-to-right shunts will eventually result in increased pulmonary arterial pressure that may alter the normal maturation of the pulmonary vascular bed [2]. The pulmonary arteriole transitions from having a reactive muscular wall to

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potentially irreversible structural changes consisting of medial hypertrophy and intimal proliferation, which manifests as increased pulmonary vascular resistance (PVR). Eventually, PVR exceeds systemic vascular resistance, leading to right-to-left shunting across the VSD, and cyanosis (Eisenmenger syndrome). The development of permanent injury to the pulmonary vessels is a function of the duration of the exposure to excessive blood flow and the anatomy.

In presence of a right-to-left shunt, oxygen-poor blood from the right side of the heart mixes with oxygen-rich blood in the left side of the heart, and the blood that is pumped out to the body is low in oxygen causing cyanosis. Cyanosis is the appearance of a blue or purple discoloration of skin and mucous membranes. It results from presence of 5.0 g/dL or greater of deoxyhemoglobin in the blood [3].

Although congenital cardiac malformations may be classified in various ways, we propose a simplified classification in four subgroups of malformations: septal defects, defects of the outflow tracts and great vessels, univentricular hearts, and other malformations.

1.1 Septal Defects

1.1.1 Atrial Septal Defects (ASD)

ASD are defects that allow interatrial communication and result from openings in the atrial septum. Defects of the atrial septum are the third most common type of congenital heart defect and the type most likely to be diagnosed in late childhood or adults (Fig. 1.1).

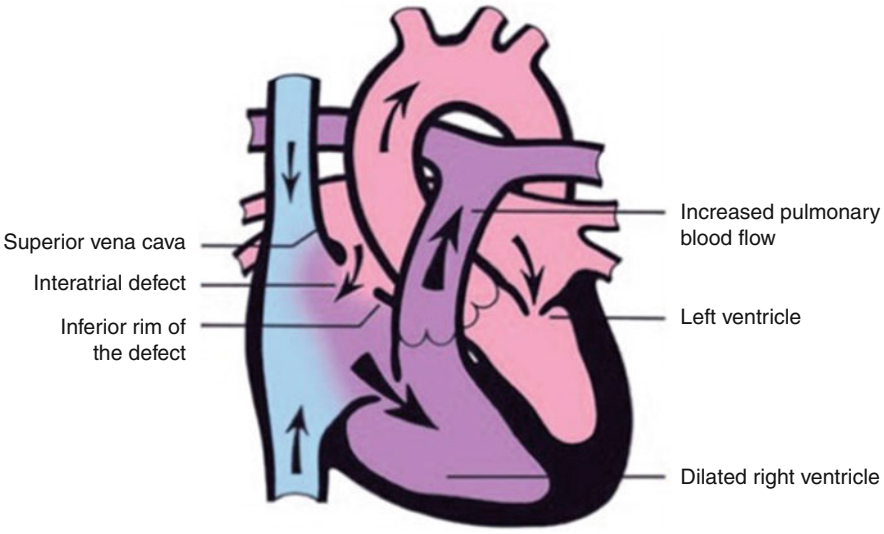

Fig. 1.1 Secundum atrial septal defect (Courtesy of the HeartLine Association)

Patent foramen ovale (PFO) is an interatrial communication that exists normally in the fetus where it allows blood to bypass the pulmonary circulation. Normally, this opening closes at birth when the lungs become functional. A PFO is seen in almost all newborns and with a decreasing frequency in older individuals [4, 5]. Complete anatomic closure of the foramen ovale occurs in 70–75 % of adults [6]. It assumes clinical importance in certain congenital heart defects and in older patients with paradoxical emboli and stroke.

Atrial septal defects are further classified into secundum and primum defects. Secundum atrial septal defect (ASD II) is a single or multiple defects within a part of the septum named septum primum. With the exception of PFO, secundum ASD is the most common cause of an atrial-level shunt. The shunt causes volume loading of the RA and RV, resulting in chamber enlargement [7]. Increased pulmonary blood flow over decades can damage the pulmonary vascular endothelium leading to an increase in pulmonary vascular resistance called pulmonary vascular obstructive disease. Most interatrial communications do not cause symptoms in childhood allowing some to go undetected until adulthood [8].

Closure of an ASD II can be accomplished by surgery or interventional catheterization [9]. Atrial arrhythmias are the most frequent late complication. Patients repaired early in life have a small risk for supraventricular tachycardia, and the risk increases with advancing age at repair. Pulmonary hypertension, another important late complication, is rare in patients operated before 25 years of age, and the risk increases with advancing age at repair [10].

1.1.2 Atrioventricular Endocardial Cushion Defect (AV Canal)

AV canal defect groups a spectrum of cardiac malformations derived from defects in the formation of the endocardial cushions.

The simplest malformation, ASD primum (ASD I) or incomplete AV canal, consists of an atrial septal defect located low in the atrial septum, adjacent to the mitral valve annulus, which is often associated with a cleft in the anterior leaflet of the mitral valve leading to mitral insufficiency (Fig. 1.2).

In other cases, the ostium primum type of defect is continuous with a larger defect in the adjacent ventricular septum. In these instances, the defect crosses both the mitral and tricuspid valvar annulae, causing deficiencies of the septal leaflets of both valves. This form of endocardial cushion defect is called complete AV canal.

Two major hemodynamic abnormalities are found. The first is the volume overload on the right atrium and right ventricle, and pulmonary overcirculation, as in patients with a left-to-right shunt at the atrial level. The second abnormality is mitral insufficiency, which leads to increased left ventricular volume because the left ventricle handles not only the normal cardiac output but also the regurgitated volume.

Infants with the complete form of AV canal frequently develop congestive cardiac failure in the first few weeks or months of life, whereas patients with ASD I may be asymptomatic, as in the ASD II.

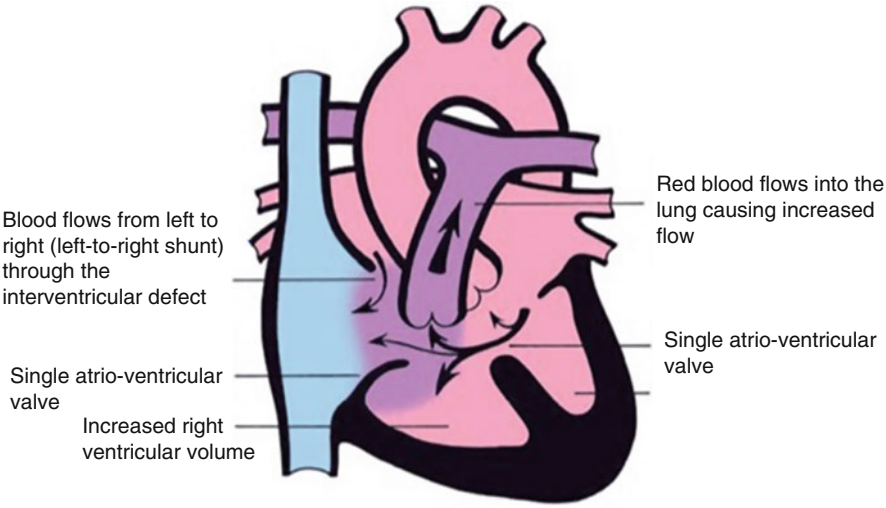

Fig. 1.2 Atrioventricular endocardial cushion defect (AV canal) (Courtesy of the HeartLine Association)

In patients with ASD I and a cleft mitral valve who are asymptomatic or who have few symptoms, corrective surgery can be delayed and can be performed at a low risk. The defect is closed; and the cleft of the mitral valve is sutured, which may greatly reduce the degree of mitral insufficiency. In patients with complete AV canal, corrective operation can be indicated in very young symptomatic infants who often respond poorly to medical management. Infants are routinely sent for operation at 2–3 months of age. The risk of pulmonary vascular disease developing within the first 6–9 months of life is high, especially in Down syndrome. The operative results are good in almost all, although some infants have such deficient anatomy of the mitral valve that prosthetic replacement of the mitral valve is required. Surgically induced AV block is likely but mostly uncommon.

Arrhythmias appear to have an earlier age of onset in patients with ASD I or repair AV canal defect than in other atrial shunts, likely due to concomitant left AV valve regurgitation. In addition, atrial arrhythmias are a common cause of deterioration [11, 12].

1.1.3 Ventricular Septal Defects (VSD)

A VSD is defined as a defect between the right and left ventricles. This defect may occur in isolation or, less commonly, may be part of a complex cardiac malformation.

The etiology of VSDs is felt to be multifactorial. Certainly, VSDs are quite prevalent in association with genetic abnormalities, especially in trisomies 13, 18, and 21 as well as other less common syndromes (Fig. 1.3).

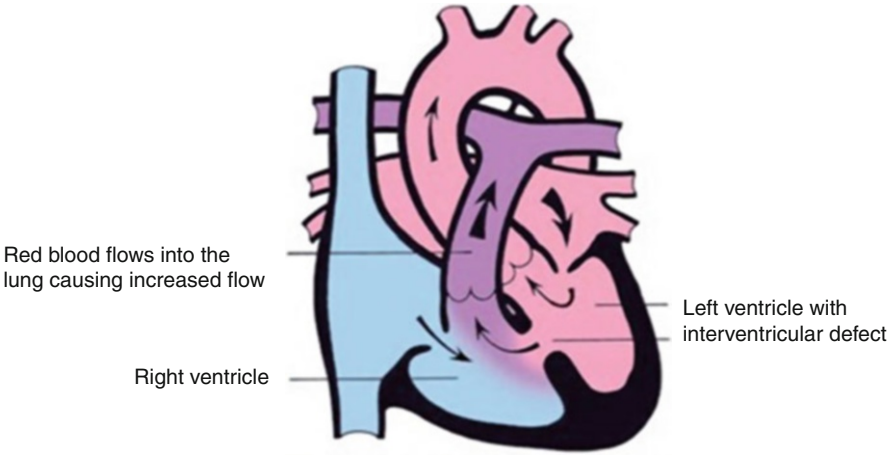

Fig. 1.3 Ventricular septal defect (Courtesy of the HeartLine Association)

Left-to-right shunting across a VSD provides greater blood flow to the pulmonary circulation than to the systemic circulation [13] leading to left ventricular volume overload and increased pulmonary blood flow. Increased pulmonary blood flow over time can damage the pulmonary vascular endothelium leading to an increase in pulmonary vascular resistance called pulmonary vascular obstructive disease. The magnitude of the left-to-right shunt depends mostly on VSD size: the bigger the defect, the larger the shunt.

Aortic regurgitation (AR) is an acquired and frequently progressive lesion in some patients with VSD [14, 15] usually due to prolapse of one or more valvular leaflets into the defect during systole.

Spontaneous closure of small VSD may occur. Closing a large VSD by open-heart surgery usually is done in infancy to prevent complications later. The transcatheter closure is a possible alternative in selected cases either in children or in adulthood.

Complete heart block secondary to injury to the conduction system during repair of a VSD may require a pacemaker in the postoperative period. The knowledge of the location of the conduction system in relationship to the defect now makes this a rare complication [16].

1.2 Defects of the Outflow Tracts and Great Vessels

1.2.1 Tetralogy of Fallot (ToF) and Pulmonary Atresia with Ventricular Septal Defect

Tetralogy of Fallot (ToF) constitutes 4–9 % of congenital heart disease and is the most common cyanotic congenital heart disease [14]. It involves four anatomic abnormalities of the heart such as (1) malaligned VSD, (2) anterior shift of the aorta

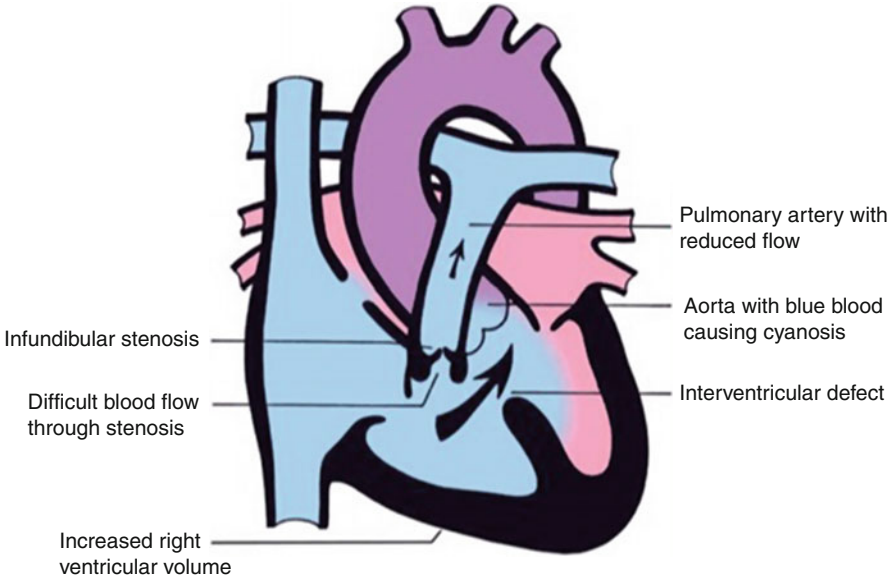

Fig. 1.4 Tetralogy of Fallot (Courtesy of the HeartLine Association)

over the VSD (overriding aorta), (3) obstruction of the right ventricular outflow tract (RVOT), and (4) right ventricular hypertrophy. Approximately 25 % have a right-sided aortic arch, and about 4 % have a coronary artery anomaly (Fig. 1.4).

The major right ventricular outflow obstruction in ToF is infundibular stenosis. The degree of cyanosis depends on the degree of right ventricular outflow obstruction. This is quite variable, from a slight obstruction to severe obstruction with pulmonary atresia. With mild pulmonary stenosis, also known as "pink tetralogy of Fallot," ToF behaves as a VSD with pulmonary overflow. As infundibular stenosis increases, progressive cyanosis due to less pulmonary blood flow develops [17]. ToF with pulmonary atresia, also known as pulmonary atresia with VSD, is a ToF severe variant [18] in which there is complete obstruction (atresia) of the right ventricular outflow tract. In this form of the disease, blood shunts completely from the right ventricle to the left where it is pumped only through the aorta (Fig. 1.5). The lungs are perfused via extensive collaterals from the systemic arteries and sometimes also via the ductus arteriosus.

Surgical treatment of the defect includes patch closure of the VSD and relief of pulmonary outflow obstruction with patch augmentation of the outflow at the expense of creation of free pulmonary regurgitation (PR). In selected cases with particular anatomic features, bypass of obstruction can be done using a right ventricle-to-pulmonary artery conduit (RV-PA conduit).

Total surgical correction can now be performed in young infants from 3 to 6 months of age or earlier [7]. Prognosis is good with total correction. After repair, the majority of patients have normal oxygen saturation and no residual shunt. The most common late complication is chronic PR. Residual RVOT obstruction and branch pulmonary artery stenosis [19] are less frequent but important late complications. Late right ventricle dilation and dysfunction are common [20].

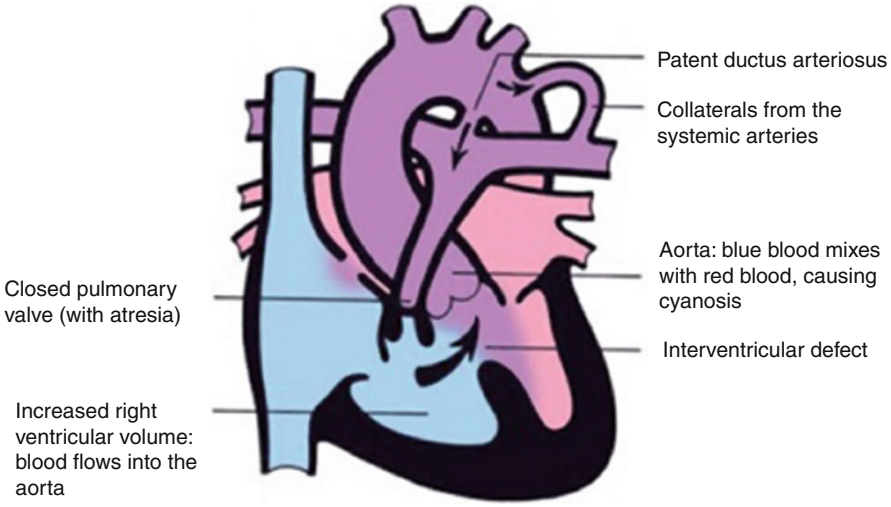

Fig. 1.5 Pulmonary atresia with ventricular septal defect (Courtesy of the HeartLine Association)

PV replacement is the treatment for chronic PR in patients with severe PR and RV dysfunction, or with symptomatic or sustained atrial or ventricular arrhythmias. Surgical valve replacement has been for a long time, the only option available for the native RV outflow tract. Nowadays, percutaneous pulmonary valve implantation (PPVI) is a valid alternative when the pulmonary valve must be changed, either in a native RVOT (in selected patients) or for a failed RV-PA conduit [21, 22]. Balloon dilation or stenting should be considered for branch pulmonary artery stenosis if flow in the artery is reduced and especially when accompanied by PR [23].

Progressive aortic regurgitation, usually associated with aortic root dilation, has been reported to occur in 15–18 % of patients [24, 25]. Arrhythmias are another important late complication. The most frequent are atrial flutter or fibrillation. Prevalence of ventricular arrhythmias increases with age and is associated with LV dysfunction [26]. Atrial arrhythmias can be addressed by a Maze procedure at the time of PV replacement [23]. Patients with documented sustained VT or aborted sudden cardiac death should receive an ICD for secondary prevention [27].

Survival after repair of TOF is less than expected for the general population at all times [28]. Sudden death due to ventricular arrhythmia is the most common cause of death after surgical repair of TOF [28]. The risk for sudden death is 3–6 % over the 25–30-year follow-up period [19].

Associated anomalies are common, such as DiGeorge syndrome.

1.2.2 Transposition of the Great Arteries (TGA)

TGA accounts for 4–5 % of all congenital heart defects [29]. Transposition derives from the Latin verb *transponere* meaning "to place across." That is, the great arteries are placed across the ventricular septum: the aorta arising from the right

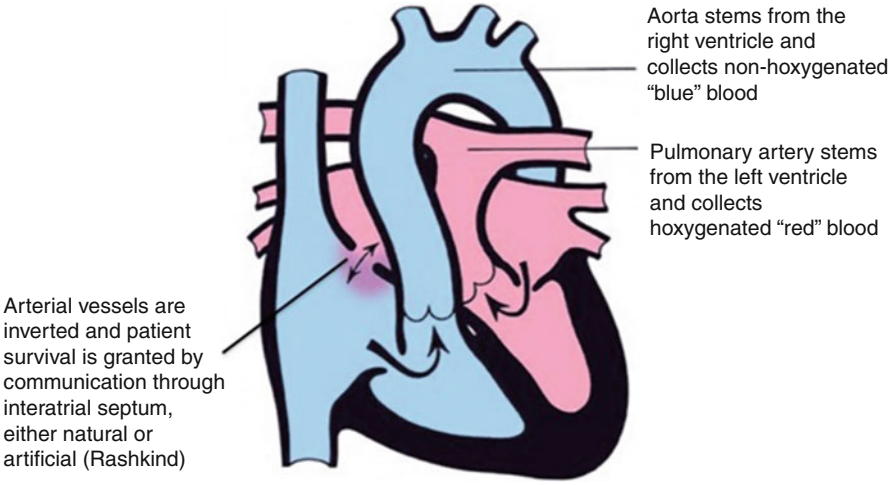

Fig. 1.6 Transposition of the great arteries (Courtesy of the HeartLine Association)

ventricle and the pulmonary arteries from the left ventricle. This is incompatible with life unless a communication exists between systemic and pulmonary circulation, as the two circulations are parallel and independent. During the newborn period, the patent ductus arteriosus (PDA) and PFO maintain this communication. As the PDA starts to close and the PFO by itself is inadequate in size, the patient develops intense cyanosis (Fig. 1.6).

At repair, the circulations are placed in series either by switching the inflow sources (atrial switch operation) or by switching the outflows (arterial switch operation and placement of a RV-PA conduit, also known as Rastelli operation). After an atrial switch operation, the right ventricle remains the systemic ventricle, while after an atrial switch operation, the left ventricle becomes the systemic ventricle. Patients born before the early 1980s most likely underwent an atrial switch operation; patients born after the late 1980s were most likely repaired using the atrial switch operation. Survival without surgery is unlikely. The arterial switch procedure offers the best prognosis with a mortality of about 5 %.

Sinus node dysfunction is the most frequent complication after an atrial switch operation. Loss of sinus node function is progressive, and by 20 years, only 40 % of patients remain in sinus rhythm [30]. Sinus node dysfunction with tachyarrhythmia or bradyarrhythmia is an indication for pacemaker therapy.

Complications related to Rastelli operation are mainly conduit obstruction and subaortic stenosis from inadequate enlargement of the VSD [31]. Several adult patients may face a right ventricular failure during their life, and a cardiac transplantation may become necessary as a treatment option.

Pulmonary artery stenosis is the most frequent complication following the arterial switch operation. Mechanisms include inadequate growth of the suture line, scarring and retraction of the material used to fill the coronary artery button sites, and tension at the anastomotic site if there is inadequate mobilization of the distal

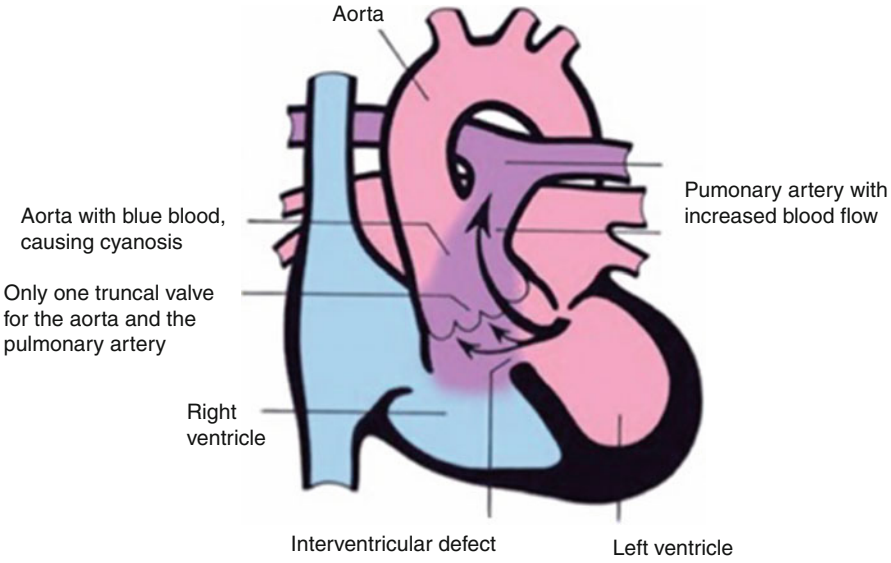

Fig. 1.7 Truncus arteriosus (Courtesy of the HeartLine Association)

pulmonary arteries [32]. Stent placement is usually effective for branch pulmonary artery stenosis.

There is a modest risk for neo-aortic valve regurgitation related in part to neo-aortic root dilatation, especially in patients with a VSD [41].

Coronary stenosis or occlusion has been discovered in 5–7 % of patients after the arterial switch operation and has been associated with ventricular dysfunction and sudden death [33].

1.2.3 Truncus Arteriosus

In truncus arteriosus, both the ascending aorta and main pulmonary artery or branch pulmonary arteries arise from a common trunk, positioned over a ventricular septal defect, that supplies systemic, coronary, and pulmonary circulations (Fig. 1.7). It accounts for about 1–4 % of the congenital heart defects [10]. Associated anomalies are common, such as DiGeorge syndrome.

Symptomatology depends upon the amount of pulmonary blood flow. With increased blood flow, symptoms of congestive heart failure develop few weeks after birth. Management consists of treatment of congestive heart failure followed by surgery. Repair of truncus arteriosus includes patch closure of the VSD so that the truncal valve is aligned solely with the left ventricle, separation of the main or branch pulmonary arteries from the truncal root, and establishment of continuity between the pulmonary arteries and the right ventricle, usually by means of a

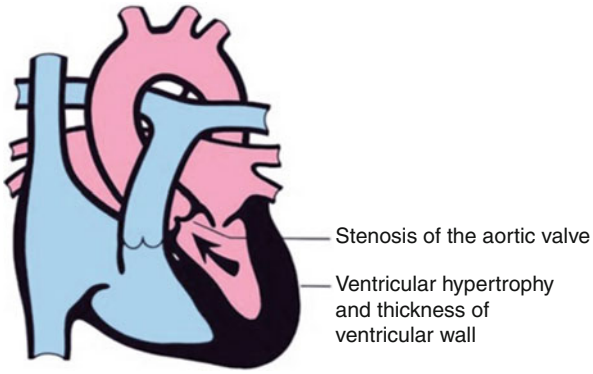

Fig. 1.8 Aortic stenosis (Courtesy of the HeartLine Association)

conduit or homograft. The prognosis is poor in untreated cases. After surgery, they will need long-term follow-up as they will eventually need to have the conduit graft replaced surgically or percutaneously.

1.2.4 Aortic Stenosis

Aortic valvar stenosis is related to either a unicuspid valve or more frequently to a congenitally bicuspid valve. The orifices of these abnormal valves are narrowed, accompanied by various degrees of aortic insufficiency in some patients. Patients with aortic stenosis are usually asymptomatic throughout childhood, even when stenosis is severe. Only 5 % of children with aortic stenosis develop congestive cardiac failure in the neonatal period, but it can develop later in childhood in patients who do not receive gradient relief. Exercise intolerance may occur. Some asymptomatic children, as they approach adolescence, may develop episodes of chest pain. These episodes signify myocardial ischemia and may precede sudden death. Syncope is another serious symptom of patients with aortic stenosis and may occur upon exercise. This symptom has also been associated with sudden death (Fig. 1.8).

Aortic valvar stenosis is progressive. Two processes probably account for this: the development of myocardial fibrosis and the decrease in size of the stenotic aortic valvar orifice by cartilaginous changes and ultimately by calcification of the valve.

Relief of the aortic stenosis gradient, either by balloon dilation or cardiac surgery, is indicated for patients with significant symptoms or for those whose catheterization data or echocardiogram indicates moderate or severe stenosis. In children, the stenotic valve is usually pliable enough for valvotomy or valvuloplasty so that an aortic valve replacement with a prosthesis or homograft is not required. Ultimately, children who have undergone aortic valvotomy may require a prosthesis or homograft in adulthood if the valve becomes calcified or rigid or, sooner, if the valve develops important insufficiency. No currently available replacement valve is perfect: mechanical prostheses are long-lived but thrombogenic, so anticoagulation is required; homograft valves, although free from thrombogenic complications, are often shorter-lived because of destruction by calcification at an unpredictable rate.

An alternative operation is the Ross autograft procedure, in which the patient's normal pulmonary valve is excised and placed in the aortic position. A homograft valve is placed in the pulmonary position, where performing balloon dilation or future surgical revision is less risky because of its more accessible anterior location and presence on the pulmonary side of the circulation. Limitations with the Ross operation are higher operative risk and longevity of the patient's native pulmonary valve functioning in the aortic position.

Subaortic stenosis is the second most common form of left ventricular outflow obstruction. This obstruction is a fibromuscular membrane with a small central orifice located in the left ventricle, usually within few centimeters of the aortic valve. A jet of blood passes through the orifice and strikes the aortic valve. The energy of the jet frequently results in alterations in the aortic valve and aortic insufficiency. Excision of the membrane is indicated in all patients to relieve the elevated left ventricular systolic pressure and to reduce the trauma to the aortic valve. The operative risk approaches that of operation for valvar aortic stenosis. The major hazard of the operation is the possibility of damage to the septal leaflet of the mitral valve, since the membrane is often attached to this leaflet. The results are generally very good, with near-normal left ventricular systolic pressure postoperatively. The subaortic membrane has a high recurrence rate after surgical correction mainly in patients operated sooner and with high peak pressure gradients, suggesting a more severe form of disease [34, 35].

Obstruction to left ventricular outflow can also result from supravalvar stenosis. In most of these patients, the ascending aorta narrows in an hourglass deformity. Although the abnormality is usually limited to the ascending aorta, other arteries, such as the brachiocephalic and even the renal arteries, may also be narrowed. Peripheral pulmonary arterial stenosis and hypoplasia may coexist and indeed represent the most important problem. The systolic pressure is elevated in the ascending aorta proximal to the obstruction. Therefore, the coronary arteries are submitted to an elevated systolic pressure that can lead to tortuosity of the coronary arteries and to premature atherosclerosis. Two factors have been implicated in the etiology of this condition. The first is Williams syndrome, in which a defect in the elastin gene is present. The second is familial supravalvar aortic stenosis, which occurs in patients who do not have Williams syndrome but they probably carry a mutated elastin gene as well. Surgery may be indicated for a lesser gradient compared with aortic valvar stenosis or if symptoms related to myocardial ischemia are present. Operative relief of the obstruction in the ascending aorta can be accomplished by surgical widening of the narrowing with a patch. Over the long term, reobstruction can occur because of progressive medial thickening of affected vessels.

1.2.5 Pulmonary Stenosis

Pulmonary stenosis occurs at three sites in the right heart outflow area: below the pulmonary valve (infundibular), at the level of the valve (valvar), or above the valve (supravalvar).

Fig. 1.9 Pulmonary stenosis (Courtesy of the HeartLine Association)

Infundibular pulmonary stenosis rarely occurs as an isolated lesion. Supravalvar stenosis or stenosis of the individual pulmonary arteries is also uncommon. In most patients, obstruction occurs at the level of the pulmonary valve.

Regardless of the anatomic type of stenosis, the right ventricular systolic pressure must increase to maintain a normal cardiac output. With the elevation of right ventricular systolic pressure, right ventricular hypertrophy develops (Fig. 1.9).

In the usual form of pulmonary stenosis, the valve cusps are fused; and the valve appears domed in systole. A small central orifice and poststenotic dilation are found. In less than 10 % of valvar pulmonary stenosis, the pulmonary valve leaflets are dysplastic: they do not show commissural fusion, the commissures are rather open, but each leaflet is greatly thickened and redundant. In most patients, dysplastic pulmonary valve is associated with various noncardiac abnormalities of Noonan and similar genetic syndromes. Critical pulmonary stenosis presents with cyanosis and failure in the neonatal period or within the first year of life. However, many of the patients are completely asymptomatic throughout childhood. Cardiac catheterization is required to perform balloon dilation, which is so far the procedure of choice for gradient relief. This procedure almost always results in a favorable outcome and reduction of right ventricular systolic pressure to normal or near normal. Even though pulmonary valvar insufficiency regularly results from valvuloplasty, it is well tolerated because pulmonary arterial pressure is low. Operative valvotomy is indicated for those patients who have failed dilation (e.g., Noonan syndrome patients with dysplastic valves) or who are not candidates for balloon dilation (e.g., the neonate with critical stenosis and an extremely hypoplastic pulmonary annulus instead requires outflow tract widening by use of a patch).

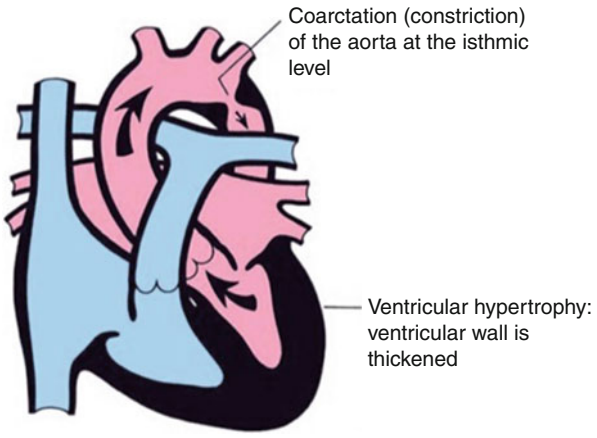

Fig. 1.10 Coarctation of the aorta (Courtesy of the HeartLine Association)

Stenosis also occurs above the pulmonary valve and in the branches of the pulmonary arteries. One or more major branches may be involved, showing either a long area of narrowing or a discrete narrowing. Peripheral pulmonary artery stenosis occurs in children with supravalvar aortic stenosis, particularly those with Williams syndrome, in patients with Alagille syndrome, or appears without apparent cause. Hypoplastic pulmonary arteries frequently accompany ToF with pulmonary valve atresia; these patients often have DiGeorge syndrome. Most patients with this condition are asymptomatic. The prognosis is extremely variable. Since the degree of stenosis is often mild and does not increase with age in most patients, it has been considered a benign condition. Apparent growth of the pulmonary arteries does occur in some; rarely, especially in Williams syndrome patients, stenosis may progress in severity. Most patients do not require surgery as the degree of stenosis is not severe. Treatment with catheter balloon dilation, sometimes with placement of endovascular metal stents, is widely used, although with variable results that depend greatly on the etiology and severity of the stenosis.

1.2.6 Coarctation of the Aorta

Coarctation of the aorta results from constriction of the tissue of the distal aortic arch at the junction with the descending aorta and near the insertion of the ductus arteriosus [36]. Often, discrete coarctation is associated with tubular hypoplasia of the isthmus and/or aortic arch (Fig. 1.10). The bicuspid aortic valve is frequently associated with aortic coarctation (85 %) [37]. Obstruction of the arch causes hypertension proximal to the obstruction and reduced blood flow and pressure distal. In adults, large collateral vessels develop that bypass the obstruction and maintain adequate resting flow to the lower body. There are several surgical techniques used to repair a coarctation of the aorta. If the coarcted segment is short and discrete, resection and end-to-end anastomosis of the proximal and distal ends is possible. If the coarctation is a long tubular obstruction, resection with interposition of a tube

graft would be necessary. In the young infant, sacrificing the left subclavian artery, and using the transected blood vessel as a graft by turning it down and sewing it into the aortic wall, was popular at one time [38]. Percutaneous balloon dilation eventually with stent positioning is indicated for recurrent and or native coarctation in adolescent and adult patients with discrete lesions [39]. Late complications include systemic hypertension, recurrence of coarctation, premature coronary atherosclerosis, stroke, aortic aneurysm formation or dissection, and endocarditis [40, 41]. Hypertension persists in up to one-third of patients after successful resolution of coarctation [42].

1.2.7 Ebstein

Ebstein anomaly is characterized by downward displacement of the septal and posterior leaflets of the tricuspid valve that are attached to the right ventricular septum. The anterior leaflet is elongated and is displaced downward within the right ventricular cavity causing "atrialization of the right ventricle" (i.e., the right ventricle is small). The "atrialized" ventricle is enlarged and thin [43–46], while the functional RV distal to the valve is variably hypoplastic [45]. The result most often is tricuspid regurgitation (TR), but in some cases, tricuspid stenosis is predominant [46].

The functional impairment of the right ventricle and regurgitation of the tricuspid valve retard forward flow of blood through the right heart. The overall effect is right atrium dilation and increased right atrial pressure, thus favoring a right-to-left shunt across the interatrial communication and/or reduced systemic cardiac output. Cyanosis depends upon the right-to-left shunt.

Associated lesions include an interatrial communication and less commonly VSD or PDA. In more complex forms of Ebstein anomaly, pulmonary valve and pulmonary artery stenosis or atresia or left-sided abnormalities such as mitral stenosis or regurgitation can be seen [47]. The conduction system is often abnormal. Transcatheter ablation is the standard treatment of an accessory pathway or other arrhythmia substrate, but success rates tend to be lower and recurrence rates higher than in the structurally normal heart [48].

While fetal or neonatal clinical presentation of Ebstein anomaly is associated with a poor outcome [49, 50], adults have a much better prognosis [51, 52]. Prognosis depends on the severity of the lesion: it is good with mild lesions and poor with severe lesions with other associated anomalies/malformations (the most frequent situation when clinical symptoms appear in neonates). Treatment is mainly palliative, and there are no good surgical options. Medical therapy of Ebstein anomaly is limited to management of complications. In older patients, tricuspid annuloplasty and rarely tricuspid valve replacement may be performed. Surgical mortality in adult patients is low, under 3 % in the current era [37]. Atrial arrhythmia is the most frequent early complication occurring in one-third of postoperative patients [54]. Recurrent hospitalizations are frequent with arrhythmia being the most common indication for readmission [54].

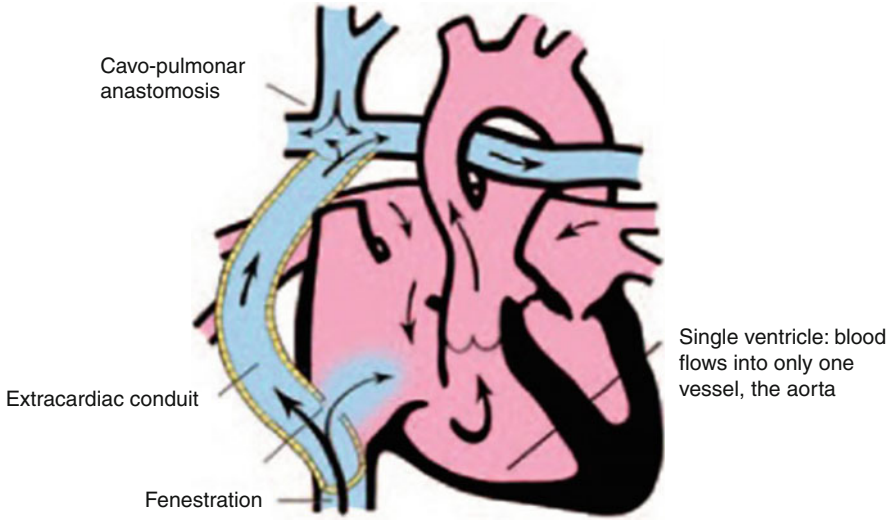

Fig. 1.11 Univentricular heart (Courtesy of the HeartLine Association)

1.2.8 Univentricular Hearts

Hearts with one functional ventricle are rare, comprising 1–2 % of all congenital heart defects. The physiology in infancy depends on several factors, including pulmonary blood flow, systemic outflow obstruction, pulmonary venous anomalies, and the atrioventricular valve function (Fig. 1.11). The Fontan procedure is the established final palliative cardiac surgical procedure in the treatment of patients with a functionally univentricular heart. In the current era, a bidirectional cavopulmonary shunt is created during infancy. Then the Fontan procedure is completed during childhood by using either a lateral tunnel or an extracardiac conduit, which is placed to redirect inferior vena caval blood to the pulmonary arteries [52]. Congenital heart defects typically staged toward this type of palliation include tricuspid or mitral atresia, double-inlet left ventricle, common AV canal with unbalanced ventricles, hypoplastic left or right heart syndrome, hypoplastic right heart syndrome, and other more rare defects.

Patients with Fontan physiology are at risk for multiple complications. Those with an atriopulmonary connection are at particularly high risk for right atrium dilation with thrombus formation and atrial arrhythmias. Both problems lead to diminished cardiac output, reduced capacity, and diminished quality of life. Heart transplantation is effective in Fontan patients with intractable arrhythmias, advanced heart failure, and protein-losing enteropathy.

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The Humanization of Healthcare Treatments and Critical Choices

2

Francesco Campione

"Humanizing" means "making human" in two ways:

(a) Making what a human being lives through human (and not brutal, inhuman, etc.)
(b) Making the ones who live, human, i.e., ensuring that individuals behave humanly with themselves and especially with others

As a result, the dignity [1] of a human being (that his/her life is given value to and is such that it is worthy to be lived) derives from his/her living experiences worthy of a human being and by his/her behaving in a manner worthy of a human being.

But what defines a "human"? Under what conditions we can consider "human" the life of a human being? When can we say that a human being is behaving in a human way towards another human being?

Being human can be defined in objective terms, in subjective terms, or in intersubjective terms.

In *objective terms*, the human can be defined as a *biological being* who, through evolution, came to possess a brain which is more evolved than all other living beings and who is the only one in possession of self-awareness and language.

In *subjective terms*, the human can be defined as a *personal being* characterized by a biography that makes him/her unique and not comparable with any other personal being.

In *intersubjective terms*, the human can be defined as a *human being*, i.e., firstly belonging to a species precisely humankind, characterized by an original and

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unsurpassed interconnection *between* all minds, all languages, all self-awarenesses, and all the biographies.

In other words:

(a) The conditions of the human being can depend on *objective* factors (the functioning of the brain, self-awareness, and the use of language that rely on it), that is, the human will be the sum of the products of the human brain, the human will correspond to objective rationality characteristics pertaining to men's thoughts, *and this is something that concerns all men and belongs to all men and also something that does not belong to anyone in an exclusive manner*.
(b) The conditions of the human being may depend on *subjective* factors (the lived biographical experience of every person), so that the human will be what each one, from his/her point of view, tells themselves they are, and everyone will be a man or a woman in their own way, and this is *something that pertains to everyone as it belongs to them*.
(c) The conditions of the human may depend on *intersubjective* factors (the meanings arising from encounters between individuals), so that the human will be the *ensemble of what the other strikes in us and affects us without belonging to us*, that which is "beyond" the objective rationality of our thought and "beyond" the particular perspective of our personal biography.

In fact:

- There are those who feel that they are men only if their brains are functioning, if they have self-awareness, and if they can express themselves; otherwise, they consider their life "inhumane" and unworthy of being lived (*being men therefore means being normal*, *functioning*, *and as biological beings belonging to a certain species*).
- There are those who feel men as long as they recognize themselves, until they feel themselves, regardless of the objective functioning of their brain, a realistic self-awareness and a normal way of expressing themselves (indeed, sometimes it's just a little bit of madness, a distorted self-awareness, and a particular way of expression that leads to a unique and unrepeatable identity). For them, life becomes inhuman and unworthy to be lived when they no longer feel themselves, whatever this means (*being men therefore means being someone distinguishable from all others*, *with a proper identity which makes one feel oneself*).
- There are those who feel men when they encounter others and feel that something about these individuals concerns them as also something about them concerns others. They feel this way regardless of the fact that others are similar or different from them when it comes to biological functioning or personal identity, i.e., irrespective of belonging to the same category of biological beings (the clever or the stupid) or personal beings (pleasant or nasty people) (*being human therefore means being there for someone else and not only for oneself*).

The truth is, of course, that these three dimensions of the human "make" man all together, but there is no concrete humanity that does not necessitate some sort of "hierarchy" to be established between them. This is so because in the absence of such a hierarchy, man could not be educated to be "human" but would become so chaotic and in a conflictual manner without any hope of becoming truly human, i.e., to organize in a harmonious unity all his/her dimensions.

In contemporary western culture, after centuries of prevalence of the personal being on the biological being and of the biological being on the human being, the hierarchy that tends to prevail is that which places the biological being before the personal being and the personal being before the human being. But the personal factor is still very strong especially in Latin subcultures.

The most evident consequence of this is the prevalence of scientific knowledge (according to which to know man, it is essential to know the functioning of the brain, neuroscience) on "humanistic" knowledge (according to which to know man, it is essential to know the feelings of each man or of mankind in general).

The consequence of this setting when it comes to healthcare is that when an individual falls ill or suffers, in my opinion, it would be desirable to promote an "ethical prevalence" of the human being on the personal being and of the personal being on the biological being.

The first consequence for those who become ill is to become a "clinical case," which can be inserted into a statistic, the only way to take into consideration their characteristics as something *that belongs to all of us and affects us all*, with an individual variability of a quantitative type with respect to a "normal" distribution in the population. In this way, the personal characteristics which make ill people unique (their personality and biography) and human (their interpersonal relationships) are pushed into the background. This is inevitably followed by a certain degree of "depersonalization" and "dehumanization." Not too much damage is done if this depersonalization and dehumanization lasts for a short time and if in this time, through the techniques of medicine, the goal of healing is reached. On the other hand, when medicine reaches its limit, and there is a failure to heal the individual, depersonalization and dehumanization can be prices that are too high to pay in terms of "loss of dignity of life" for patients who base their value on the "personal feeling of being themselves and unique" and on those who base it "on the consideration of others."

The prices to be paid and the manner in which they are paid have been outlined previously in the "Manifesto for the humanization of medicine" [2]:

> …In sickness, we all live the *insult* to the integrity of our body (integrity that is the basis of the possibility to build a personal history) and the *humiliation* in the pride to exist (pride that is the basis of the ability to respect and to be respected). And when we are ill, we all invoke a medicine that has the power to heal, to alleviate pain and to prevent the threat of death. But medicine is not able to counteract this offense and this humiliation, if not in the rare cases in which it helps us to achieve a complete and lasting healing. This is so because, in order to help us, Medicine is in possession only of its technical power. As if to say that to overcome the offense that disease incurs to the dignity of the person and the humiliation to the dignity of man, Medicine possesses an uncertain and therefore anonymous statistical

> power to restore health. However, it is the insult and humiliation of the illness that the uncertain power of the doctor is unable to effectively overcome, because it almost never heals permanently. This injury and humiliation will be able to be counteracted if Medicine will give the offended person and the humiliated humanity of the ill a moral value higher both than that of the personal being and of the humanity of the healthy person and also of Medicine itself. In other words, the doctor should recognize the *moral majesty* of the ill precisely in the offense, in the humiliation that the disease inflicts, therefore putting his/her technique to the service of the sick person's dignity as a person and as a man!
>
> But what gives dignity to the sick, as a person and because they belong to the human race? This is what every human being shares with all other human beings and that what gives it meaning with his/her personal differences but it also goes beyond these differences. This is about the possibility that every sick man has to stir in another human being the responsibility of taking care of him/her by asking him/her for help.

It then becomes important to *empathize* with patients in order to be inspired by them (allow them to tell) to do and say what could "compensate" the loss of dignity of life determined by depersonalization and dehumanization. It is a concrete operation of appreciation; it refers to everything that gives value to each one of us as a human being, regardless of our biology and of us "being a person." Simone Weil [3] stated that the value of each person, whoever he or she is, is based on their "desire for good." This is what was described in the cited "Manifesto for the humanization of medicine" [2] in the following manner: "It is important to ensure that the moral superiority of the ill (compared to those who assist them) is also recognized by the ill people themselves, placing as a focal point of the doctor's care the restoration of the ill person's dignity even when (and especially when) it is not possible to achieve this objective through healing. For this to occur, it is essential to ensure that the training of doctors (and the entire medical staff) is no longer predominantly technical and that they are no longer blackmailed economically but that they are trained to become what they always have been and what they always wanted to be in their most authentic and most noble vocation: the guardian of life and the dignity of the ill person in spite of any economic compatibility and kind of power whatsoever. It is also important to aid those who are momentarily healthy and gain awareness that when the offense and humiliation of disease occur, for everyone sooner or later, it is possible to adopt more effective defensive strategies than medical technologies: the sharing of a collective moral principle that those who suffer " are worth more morally" than those who do not suffer or those who suffer for them, a principle that can inspire the responsibility of a doctor capable of a technical act which is not self-sufficient but always at the service of the sick person and the humanity of the ill person."

It is not difficult to articulate what has been said so far in terms of practical assistance as long as doctors do not find themselves facing "choices" that may cause them distress (basically, when and to what extent to refrain from active therapies and when to limit themselves to supportive care or palliative care) [4].

In this respect, the example of "intensive therapy" is the clearest and most "inclusive" example, given that in all diseases that are life threatening, when the situation is aggravated, it must be decided if there is a need for "intensive therapy" and, if so, up to what extent it should be carried out.

The intensive therapy units are among the healthcare places in which more frequently and dramatically there is the problem of what is the "right thing" for the ill person, due to the difficult choices (both therapeutic and not therapeutic) which are imposed every time there is a comparison of different interests, different knowledge, different sensibilities, and different ethics on life and death. *Humanizing* intensive therapies means (as in every healthcare context) during the first attempt to manage gradually to "do as much good as is possible" for everyone (patients, caregivers, and healthcare providers): both in the cases of a "lesser" choice like making the departments more accessible or leaving them closed to the family members as they most often are to this day and age and also in the cases of "greater" choices, such as the interruption of therapies (both artificial and not), to let patients die without hope or to continue to provide care fostering hope which is more or less illusory.

Perhaps rightly so, or perhaps because in our culture it cannot be done otherwise, when defining the "good" which humanizes therapeutic choices, we tend to repropose also in medicine the model which *Plato* originally presented in "Filebo" [5], one of the dialogues which is not very well known. In this dialogue, Socrates searches, together with friends, the "true good" for mankind. In summary, the conclusion Plato reaches is the following: the true good for man is the happiness (or pleasure) in life to which the truth must be added. Without this truth, any happiness would become false; the supreme good is the "proportion" that makes the mixture of pleasure and knowledge become "good," and this composition is always necessary in order to obtain goodness.

What also could we intend saying when talking about "proportioned care," if not that it should be a proportioned mixture of interventions which really increase the well-being of everyone, that is, it makes others feel better and that it is truthful?

If anything, this issue, already present during the times of Plato, refers to whether the proportionality is obtained "ethically" through a virtue which he intended more or less as being wisdom, moderation, alleviation, or equilibrium, or if it is obtained through mathematical principles and their application of the phenomenological field.

This depends on the reply given to the following question: *the medical care needs to be proportionate*, *but proportionate to what*?

In our culture, there has been a dispute between two fields which has lasted some centuries, between two criteria to "proportion" medical care: the biological criteria and the personal criteria. Here they are in detail:

I. From the point of view of the biological knowledge, medical care is proportionate if it is commensurated to its end and it results as being effective; it is not proportionate if it is not efficient when it comes to its finality. For example, feeding a patient who is in an irreversible coma is "proportionate" if it is done to keep him/her alive and it is "not proportionate" if it is done to make him/her come out of the coma. But who decides the finality of an interventional medical treatment? This is precisely the limit of this criterion: it is valid *if* no subjective aspects interfere with the certainty of the objective finality of the medical care

(when it comes to this example, up to the point when someone does not want to use artificial nutrition not to keep the patient in the coma alive as a scope in itself, but to consent that a cure is discovered in order to awake the patient from the coma). However, the value of the humanization of the biological proportionality needs to remain present in order for goodness to be produced for everyone, whenever there is a medical care, in which there is a shared finality and which is also efficient in improving the objective condition of patient and as a consequence of the caregivers and healthcare providers.

II. On the other hand, from the point of view of the awareness, that is, the intentionality of the people (*patients*, *caregivers*, and *healthcare providers*), a cure is proportioned if commensurated to the way these subjects are aware of the conditions (whether they accept them or they deny them) and their expectations. For example, feeding a patient who is in an irreversible coma is "proportioned" for who accepts the condition of life in a coma (either because there is an instrumental belief that the patient might wake up the day after or because the same value is continued to be attributed to the life of the patient in coma); on the other hand, it is "not proportioned" for those who refuse the condition of life in a coma and who desire that this life ends as soon as possible. But who is right? Both, from the subjective point of view until they remain "separate" in the private life. The limit of this personal criterion when it comes to the choice of healthcare consists precisely in this: it is an individual criterion which requires a "pluralistic" community, made up of individuals with different rights and duties, that is, with different subcultures which need to be very tolerant between themselves and also kept rigorously separate; otherwise, they will be in constant "civil war." Also in this case, however, there remains the value of humanization of the personal proportionality of healthcare, since benefits are produced for everyone every time healthcare is commensurated to the people's reaction and to their expectations, and it is provided in such a way "not to hurt anyone else."

In summary:

(a) The proportionality of healthcare, in the way it has developed in our culture, is made possible, on the one hand, by the ever-developing capacity of objective science to measure the *efficacy* of healthcare, measuring them to a standard, and on the other hand, by the capacity to assist and to allow to emerge the intentionality and the expectations of the ones receiving healthcare in an empathic manner, by commensurating the healthcare provided.
(b) The limits of the proportionality of healthcare are, on the one hand, dependent on the limits of science in predicting their own developments and, on the other hand, by the irreducible conflicts of the different personal points of view.

Therefore, a "sense of proportions" needs to be developed, by following the platonic indications of wisdom, a "sense of proportions" both on behalf of science (in a way in which one should proceed with moderation, prudence, and humility) and also on behalf of the single people (in a way that they should act in a poised and

tolerant way towards others when they have a different point of view, making sure that the civil war between different points of view is not external and irreducible).

Will this suffice? And *if* it shouldn't suffice, are there any other alternatives?

In order to discover this, we must ask ourselves if the *human criteria* can show us another way, different from the ones which are proper of the two ways of pursuing well-being illustrated previously.

The biological healthcare humanizes the existential conditions of the actors of the intensive therapy units (or of healthcare places of this kind) through their objective efficacy in making patients feel better or less worse, that is, bringing "in themselves" their beneficial value, increasing the *well-being of everyone and of nobody* in particular, *the well-being on average*.

Care on a personal level humanizes to the point that it corresponds to the subjective manner of becoming aware (by accepting or refusing) the need to receive healthcare and to the degree of realizing the corresponding expectations, that is, to what extent well-being is identified in that which is *good "for someone."*

The human dimension characterized by identifying what is good in what *is good for everyone and for each one at the same time* is clearly left out. And it is precisely by taking into consideration this third dimension that the pursuing of good can be seen as being different both from the biological and personal dimensions. In other terms, the good can be pursued through "human healthcare" that (instead of being based on efficiency that corresponds to the satisfaction of objective needs or the adherence of the intentionality to be treated of the single patients, which corresponds to the satisfaction of the unique and non-repeatable needs of a single person) are based on the concept of taking on ourselves the *responsibility* "for all the others" of their own good, first and foremost and irrespectively from having the availability of objective and adequate means or knowing subjective ways of becoming aware (accepting or refusing) of situations and expectations which are relative to them. This is about a human dimension, which in our culture is neglected also on a theoretical point of view. This is so in spite of the fact that these healthcare situations in which someone pursues the well-being of another person, and therefore humanizes the situation, taking care not only of his/her needs but also of the needs of everybody else, without knowing if there will be the "power" to act and "before" knowing who is the other person being helped as a person. It is true as Severino [6] states in one of his writings that nobody knows what is the absolute good, but it is clear to everyone that good which is so in the indifference for everyone or only for someone cannot be called as "good" whatever it is. Thus, a dimension of the good appears which is irrespective of its "what" is, and it consents us not to turn away from looking for the good, with the excuse that it is not possible for us to understand its essence [1].

[1] For now, we must resign ourselves to what has always happened: the winning force has been called truth, justice, and law. For this reason, it is pathetic to invoke an absolute good. Nowadays, the dominant culture is not capable to resolve these problems. They are resolved in a practical, political manner.

This is a criterion of humanization in which there is no concern of commensurating healthcare to the standards of efficiency and not even of prorating it to the "who" is the single recipient, but it is based on something which comes "before" (the responsibility that one feels who is committed to provide healthcare) and is incommensurable (consisting in the commitment of considering the points of view of all the others without seeing what is convenient, that is, adopting an attitude which is "without calculating" of someone who is committed to do good for others in a disinterested way, by being disinterested about what extent he/she will be able to do it and without knowing the first intentionality and expectations of the person to whom the good is being done). This is the human dimension which contradicts the platonic model of the "true good," dethroning the value of measure and proportion, both when there is *calculation* and *quantification* and also when there is *qualification* and *personalization*.

It is a humanization that is completely different from the Promethean Humanism (that consists in giving man "false hopes" making him ignore "when" death will arrive) and from the Christian one (which consists in giving man the hope of a life after death) which gave rise to the crises in our period.

The Promethean Humanism fails because it does not manage to keep together the two gifts that Prometheus makes to men: fire and the false hopes based on the ignorance of time of death. The "fire" of the proportionate and technically efficient healthcare has in fact become incompatible with the false hopes of therapeutic persecution.

Also Christian humanism fails as it tries to validate the false Promethean hopes in a perspective focalizing on new technological progress, instead of residing faith in a redeeming God, who is perhaps already dead.

Only the imbalance in responsibility, which is based on placing before oneself the other and everyone else (aiming to conciliate the good of everyone and each one), could allow us to overcome this block every time the two proportionalities possible (the technical-biological and the personal one) encounter their limit.

What does this mean concretely/in actual terms?

First of all, it means that every time science ignores the "when" of its efficacy, we can still humanize the situation of those who suffer (i.e., pursue their well-being) taking on just the same the responsibility to help them "before" knowing what and when we will be able to objectively do so. For example, we cannot scientifically predict when we will be able to make someone wake up from a persistent vegetative state, but we can still help those who need to do it, taking on the responsibility to help them and thus getting involved in their "infinite desire" to make them wake up and involve them in the "study" without time limits (the necessary time that it will take) to reach this result.

Secondly, it means that every time a person does not accept his/her condition or would like something that is impossible in order to hope to accept it, we can still humanize the situation by still taking on the responsibility of helping them, even though we do not know if we will be able to meet their expectations.

For example, if the doctors who were taking care of Eluana Englaro [2, 7] had taken on the responsibility of her father's request to make her die, instead of

defending themselves behind their technical duties, denying him help, and even condemning him morally for this request of making her die, the human involvement which would have been determined "between" them would have spontaneously limited the will of Beppino Englaro, making possible to allow a dialogue which was previously not possible, between the two contrasting violences of causing death and forcing to live [2].

In other words, this "humanism of disproportionate responsibility" will be able to unlock a new possibility: *to pursue the good of everyone and of each one*, which does not mean making a calculated average of what is objectively available and not even favoring the particular good of who imposes himself/herself on others (by using some force or with the power of complaint), but rather sharing, all of us, the responsibility of doing the "right thing" for everyone, continuing in this search for good in infinity even after having done what could have been done in the given circumstance. In other terms, what can be done objectively and also what the interpersonal situation imposes that needs to be done, will be done, but in the perspective of the infinite research of "justice" for everyone, that is the good for everyone and for each one.

An example.

The healthcare provided needs to be proportionate (in the two senses indicated above) even in the situations of clinical irreversibility (such as the persistent vegetative state or *terminality*), but one needs to be able to call into question the choices made every time that new knowledge changes the standards of care and a subject reacts in a particular way. And we can clearly manage to do so only if the time of efficiency (the time of the standard good) and the personal one (*the time of the subjective good*) are inscribed in an infinite time (*the time of the good for everyone and for each one*, the necessary time, i.e., the time of desire). By pursuing in infinity the "right thing," we can sustain the damage derived from the inevitable limits of the choices based on the objective awareness which is always partial and those based on individual preferences which are not valid for everyone.

Expressing in more simple terms all that has been said up to here, we can say, reaffirming what we are trying to say in this chapter, that the *human* pursued in the humanization can be defined in objective, subjective, and intersubjective terms.

In objective terms, the human being can be defined as a *biological being* who, thanks to evolution, uniquely possesses a more evolved brain when compared to all other living beings, self-awareness, and language.

[2] The Englaro case is, in some respects, similar to the Terri Schiavo case, only with a less conflictual family. Eluana lived in a vegetative state for 17 years after a car accident, until her death caused by the interruption of the artificial ventilation. Eluana's father, Beppino Englaro, had tried for 17 years to get recognized to the Italian law the right of his daughter not to be kept alive artificially and therefore to be allowed to die, arguing that Eluana would not have wanted to live in a vegetative state. Since there was no biological testament, Beppino Englaro made himself guarantor of the will of his daughter together with his wife and he sought to corroborate this position through the testimonies of the friends of his daughter.

In subjective terms, the human being can be defined as a *personal being*, characterized by a biography which makes him/her unique and not comparable to any other personal being.

In intersubjective terms, the human being can be defined as human, that is, first of all belonging to a species, precisely the human one, characterized by a primary and insuperable interconnection between all minds, all languages, all self-awarenesses, and all biographies.

Naturally, these three dimensions of the human "make" man all of them three together, but there is no concrete humanity which does not necessitate to establish some kind of "hierarchy" between them.

After centuries of prevalence of the personal being on the biological one and of the biological on the human one in western contemporary culture, the tendency which tends to prevail is the one which places the biological being before the personal one and the personal one before the human one. But the personal factor remains extremely strong especially in the Latin subcultures.

The consequence of this when it comes to intensive therapy units is that the ill, since they have been profoundly affected in their biology by illness and traumas (mostly justifying in this way the hierarchy of the human which makes the biological dimension prevail), more than in any other healthcare context, have the risk of being "reduced" to biological beings. This means that their condition is humanized only from a biological perspective. For this reason, when "humanizing" care for human beings, it is necessary to take into consideration: (1) his/her personal dimension (what makes him/her feel like a person, his/her wishes and the ways of telling himself/herself what is happening) and (2) his/her human dimension (what makes him/her feel "one like all others," on the basis of the value attributed to him/her as a person despite the impairments sustained).

Naturally, the same partial humanization of healthcare can also interest the family members of the patient and the healthcare personnel when there is a tendency to disregard the human dimensions in the interventions. More specifically:

(a) The *intensive therapy* unit care is "humanized" for the family of the patients (other than indirectly improving the biological conditions of the ill): when they are helped to overcome stress and the traumas which can be inflicted in a highly stressful and traumatizing environment; when they are helped to personalize their participation to life in an intensive therapy unit allowing them, as much as possible, to choose how and when they visit, so that they feel as much as possible at home and thus guaranteeing the maximum collaboration on assistance; and when a positive moral value is attributed to whichever request is made from the family members, in order to establish with them an empathic communication based on a respectful welcoming of the request in itself. In this manner, the impression is given that the staff which assist support whichever request and it is possible to ask for something even when it is not allowed.
(b) The intensive therapy unit care is "humanized" for the healthcare personnel (besides improvements through objective professional training): when they are helped to deal with feelings of inadequacy and helplessness of who does not

manage to do what they should without being indifferent, an *indifference* which makes them suffer less but which can lead to a more or less prominent abandoning of the patients and their family resulting in a dehumanization of the condition; when they are helped to appreciate the uniqueness of the patients who cannot express themselves or to personalize the participation of family members in the daily life of the unit, without identifying themselves to the point of behaving *as if* they themselves were in their place; and when they are helped to attribute a *superior moral value* to the patients in order to make them feel human despite of their biological and personal conditions and to establish an empathic communication with their family, in which there is the ability to respect their requests even when it is not possible to fulfill them. Finally, also not to regress to a personal concern or a technical indifference in the cases, they discover themselves as being not sufficiently disinterested in the help of the patients and/or their family.

This is the beginning of the "human" pursuit to ensure that this evermore broad and complete humanization is possible in its concrete dimensions.

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Part II

Psychological Aspects and Neuropsychological Functioning in Children and Adolescents with ConHD

Psychological Aspects in Children and Adolescents with Congenital Heart Disease and Their Parents

3

Elisabeth M. Utens and Eveline Levert

3.1 Introduction

Children with congenital heart disease (ConHD) constitute a heterogeneous group, in which mild and severe cardiac diagnoses are represented. The dramatic improvements of medical and surgical treatment for children with ConHD have led to increased survival rates during the last decades, also for children with the most severe conditions. Whereas in former days, about 85 % of the children born with ConHD died, nowadays about 90 % of the children with ConHD survive into adulthood [1].

Due to this hugely improved survival rate, the focus has shifted from mortality to morbidity. Children with ConHD may have cardiac residua and sequelae after surgical or interventional treatment, and lifelong medical follow-up is recommended [2]. With the focus on morbidity and ConHD now being considered more as a chronic condition, this has put forward the question: "What is the psychosocial impact of living with a congenital heart disease, both for parents and their children and the parent–child interaction?"

The aim of this chapter is to give an overall outline of psychosocial aspects and problem areas, related to living with a ConHD, both for parents and their children, for different life phases and the parent–child interaction. Following the chronological order form birth to adulthood, we will discuss psychosocial aspects and problems for:

1. Parents of children with ConHD
2. The parent–child relationship

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3. Preschoolers and school-age children with ConHD
4. Adolescents and transition-related topics

In these four parts, the following aspects related to living with a ConHD will be addressed: (a) emotional/behavioral problems, (b) intellectual and academic functioning, (c) quality of life, (d) motoric functioning, and (e) social functioning. Finally, a clinical case will be presented.

This chapter does not aim to give a complete review, but rather focus on psychological areas, where a psychologist could provide help and counseling attuned to the needs of the individual child and adolescent with ConHD and the parents.

Each of the four parts will be ended by conclusions and practical implications.

3.2 Psychosocial Aspects for Parents of Children with ConHD [3]

3.2.1 Learning the Diagnosis [3]

When parents learn that their child is diagnosed with a congenital heart disease (ConHD), they may experience overwhelming emotions and intense stress. For all parents, the birth of an infant is a very emotional, important life event that brings along a mixture of feelings of joy and excitement but also increased stress.

For parents of ConHD children, the normal transition to parenthood is burdened by the extra stress of having to adjust to this overwhelming situation, the fear of an uncertain future for their child and the demands of learning how to take care of their baby. The heart has the symbolic meaning of life. With the diagnosis, questions as "how long will my child live, will he/she will healthy, how will the future be, and how can I take care of the baby?" immediately come to mind. Parents often state they feel as if the "ground was swept away beneath their feet" and that "their world fell into pieces" when they learned the diagnosis.

In taking care of their infant with ConHD, parents may have to face:

- *Physical demands* (such as problems feeding the infant)
- *Financial costs* (medical and transportation costs, lost income)
- *Practical problems* (day care for siblings, combining care with work, taking days off from work for fathers)

3.2.2 Parental Working Through and Coping: Short Term [3]

In this overwhelming and confusing period, parents mostly experience a process of "working through." This can be compared to a mourning process: parents mourn the loss of their expected and "fantasized healthy baby" [4]. They may experience a broad range of feelings: grief, anger, sadness, resentment, guilt, uncertainty, helplessness, and hopelessness.

This process takes time and is a normal and natural reaction of parents to an extremely stressful situation. The length and intensity of this "working through" process varies between parents and may be imbalanced between marital partners. The differences in the working through process between the individual partners as such do not reflect the quality of the marital relationship. It may simply mean that partners may have a different way of adjusting and different way of coping [3].

How parents cope with the situation and their emotions is influenced by their:

- Personality characteristics (e.g., parental mental health and coping prior to the birth of the ConHD child)
- Relationship with their partner (Happy parental relationship? How well do parents communicate? Can they support each other?)
- Social support networks
- Other (healthy?) siblings in the family
- Socioeconomic status
- Financial and work situation
- Knowledge of congenital heart disease

In contrast to what might be assumed, Lawoko and Soares [5] reported that parental reactions and stress are *not* related to the severity of the cardiac defect as such. Factors such as financial burden, time needed to care for the ConHD child, distress, and social isolation, rather than the severity of the disease, were related to reduced quality of life in parents of ConHD children.

Taking care of a chronic somatically ill child has been described as one of the most stressful experiences for any family. Moreover, it has been reported that parents of children newly diagnosed with ConHD report relatively more stress levels than parents of children newly diagnosed with other chronic illnesses [6]. Noteworthy, these overwhelming emotions are commonly experienced at short term.

3.2.3 Parental Working Through and Coping: Long Term

On the longer term, however, parental stress and coping may change and are influenced by the course of the illness. In the Netherlands a cross-sectional study was conducted in a cohort of children, at least 7 years after their first surgery for ConHD [7]. Remarkably, this cohort study showed that on the long term, parents of these ConHD children report a *better* mental health (less distress, less *somatic* symptoms, less anxiety, sleeplessness, or serious depression) and more adequate means of coping than parents of healthy children from the general population. These findings indicate that parents may develop other norms and values once the stressful period of the cardiac diagnosis and surgery has passed. Their experiences may have made parents strong and led them to worry less about the futilities in life. This phenomenon is also known as "posttraumatic growth" [8]. Posttraumatic growth indicates that a traumatized person may experience a positive psychological change due to their very stressful negative experiences [8].

Noteworthy, in the Dutch cross-sectional study described above, the long-term medical state of most patients was stable, and most patients did not need rehospitalizations. However, in times of new hospitalizations, invasive procedures, and renewed crises, parents will have to go through an adjustment phase again.

3.2.4 Conclusion, Clinical Implications Regarding Parents of ConHD [3]

In conclusion, if parents learn that their child has a ConHD, they may experience overwhelming emotions and intense stress, which can be considered a natural reaction to an exceptional stressing period. Parents undergo a process of "working through" and adjusting to the situation, which takes time and is different for each individual parent.

At long term, when their child's condition is stable, parents have been reported to have a better mental health compared to the general population [7].

3.2.4.1 Clinical Implications

First of all, clinical psychologists can support parents by listening in an empathic way, providing parents the opportunity to express their feelings and tell their narrative stories. Psychologists can provide psychoeducation: explaining to parents it is important to accept their feelings and realize that their emotions, however extreme, are a normal part of the coping process.

It is important that parents express their feelings; share their worries, fears, and hopes; and accept their limitations. Moreover, it is important that parents do not blame themselves for not being super parents (e.g., if a parent feels too anxious to accompany the child to the operating room, then it is better to express this, so that the other parent, a grandparent, or a nurse can take over).

In our opinion, not every parent needs the consultation of a psychologist when feeling depressed, angry, or very anxious, since these overwhelming feelings are part of the normal adjustment process. Formal counseling by a psychologist is recommended when [3]:

- Parents request it.
- The parents' feelings are overwhelming and out of proportion to the "normal adjustment phase."
- Parents get stuck in the process of adjusting and coping.
- There are difficult social and family circumstances.
- There are additional psychological problems.

3.2.5 Psychosocial Aspects of the Parent–ConHD Child Relationship

The presence of a cardiac defect may alter and complicate parental attitudes towards children, irrespective of the severity of the congenital cardiac malformation. Early

difficulties with feeding or care giving may harm the parents–infant relationship and especially the mother–child attachment. How parents adjust to and cope with the entire situation (diagnosis, hospitalization, surgery, invasive procedures) and also parenting their child at home can negatively influence both the short- and long-term development outcomes of the ConHD children [9].

Studies into parents of ConHD children of all ages have shown that these parents are vulnerable in the development of establishing "normal relationships" with their children. Family variables such as parenting style, marital status, and maternal mental health difficulties (maternal anxiety and distress) have been proven to be more powerful predictors of unfavorable behavioral outcomes in ConHD children than disease factors, such as severity of disease [9].

As to the *perceived severity of* the cardiac defect, both underestimating and overestimating the severity of the cardiac defect can have a negative influence on the development of the child [3]. Underestimation may lead to noncompliance with hospital checkups or asking too much of a child (e.g., a physical education teacher underestimating the ConHD and requesting an adolescent to perform the Cooper running test in time and giving the teenager a poor grade). Overestimation may result in overprotection and reduced physical activities (sports and leisure time activities), which may increase social isolation, loneliness, and depression in the child [3]. Inadequate *knowledge* about disease severity may lead to misperception of disease severity. From international literature it is well known that parents have important knowledge gaps as to the cardiac defect of their child [10].

From clinical experience it is well known that parents often report that the time of diagnosis and surgery was too stressful for them to absorb information and that they felt overloaded with information. Due to time pressure during outpatient clinics, parents may feel embarrassed or unable to ask questions. Furthermore, communication should be attuned to the intellectual abilities of parents, by giving repetition, time to assimilate the information, and understandable reading materials.

A special remark note: parents of children with mild defects have been reported to be at risk of receiving less attention from the medical doctors. Since children with mild ConHD have less frequent outpatient examinations, they may receive less information compared to children with more complex ConHD [11]. Since these parents may also have misperceptions regarding the disease severity, it is possible that they might limit their children when unnecessary. It is therefore recommendable to also spend necessary time in educating parents of children with milder ConHD.

3.2.6 Conclusion, Clinical Implications Regarding the Parent–ConHD Child Relationship

3.2.6.1 Conclusion

The presence of a cardiac defect complicates parental attitudes towards children, irrespective of the severity of the cardiac defect [9]. Parents have shown important knowledge gaps as to the ConHD of their child [10]. This may result in misperceptions as to the disease severity, which may harm the child's emotional development.

3.2.6.2 Clinical Implications

There are several protective factors when it comes to the parent–child relationship, and it is important to foster them in order to positively influence the emotional development of ConHD children. Psychologists can facilitate this process by psychoeducation. This includes explaining to the parents that the most powerful, beneficial, healing, and protective factor for their child is to love and accept their children just as they are and to try to educate them as normally as possible [3]. From a clinical psychological perspective, it is crucial that a child feels he/she is unconditionally loved and accepted by his/her parents. In this context, Cohen et al. [12] found that ConHD adolescents reported higher perceived parental acceptance than healthy adolescents. Parental acceptance had a stronger influence on the psychological well-being of these adolescents than parental control (which was perceived in fact as lower compared to that in healthy controls).

Other protective parenting styles may be stimulating autonomy, physical and social activities, networks, and/or compensatory activities in children with ConHD. These factors may foster self-esteem and emotional resilience. These topics will be discussed in the next paragraphs on children and adolescents with ConHD.

3.3 Psychosocial Aspects of Preschoolers and School-Age Children with ConHD

3.3.1 Psychosocial Aspects of Preschoolers with ConHD

Toddlerhood is also known as the separation–individuation phase [4]. Toddlers are preoccupied with differentiating themselves from others, and they are looking for predictable effects of their behavior on others. They are typically absorbed by ritualism (making things predictable) and negativism (gaining control over the environment). Moreover, they strive for autonomy [4].

Toddlers may experience medical procedures or hospitalizations as anger from the parent or loss of parental love. They may react to hospitalizations by passive and depressive behavior, but also anger. The separation from parents is a sensitive and challenging developmental task during toddlerhood. As a reaction on the separation from parents, the so-called “regressive” reactions are well known (e.g., a toddler who has successfully finished toilet training may wet his diapers again after a hospitalization) [4]. Possibilities for “rooming in” can help to prevent such regressive reactions and post-procedural psychosocial problems for the child.

Preschoolers (4–5-year-olds) overall may cope better with being separated from their parents. Due to magical thinking, which is typical for their developmental phase, they may have fantasies of mutilation or castration regarding invasive medical procedures. Therefore careful and proper educating the child prior to procedures is essential.

During hospitalizations parents may pamper the child or “spoil” him/her to a certain extent. Then, after dismissal from the hospital and back home, it may be

difficult for parents to set limits. Parents may be inclined to pamper their child, due to a natural reaction to be protective as much as possible, since the child had to undergo such painful procedures. They may suffer from feelings of guilt that the child had to undergo all this, and as a consequence they may have difficulty in disciplining their child. Toddlers with ConHD may show temper tantrums just as any healthy child. Moreover ConHD toddlers may behave angrily and in an irritated way after hospitalizations. Parents may then worry: "is it bad for the patch in my child's heart if he/she cries and screams so firmly?" A parent may therefore give in to the child, out of anxiety. Moreover, after such a stressful period, parents may feel exhausted and may therefore also be inclined to give in (go the easy way).

Through clinical experience it has been observed that toddlers may show eating problems, separation problems, and sleeping problems (nightmares) following hospitalizations. These reactions may last several months, half a year, or longer.

In a Dutch sample of 2–3-year-old children awaiting elective cardiac surgery or catheter intervention, increased levels of total emotional and behavioral problems were found, compared to normative data [13].

3.3.1.1 Clinical Implications for Toddlers and Preschoolers with ConHD

Psychologists may support parents in the complex and difficult task between providing a feeling of safety and warmth for their child and setting limits and disciplining their child. A key principle in cognitive behavioral therapy is that parental reinforcement of unwanted behavior in the child (e.g., screaming, angry behavior) may result in the child going on screaming also in the future. Reinforcing and stimulating desirable and wanted behaviors is more beneficial for the child's psychosocial development. For further clinical implications, we refer to paragraph 3.2.5 of this chapter.

3.3.2 Psychosocial Aspects of School-Age Children with ConHD

3.3.2.1 Behavioral and Emotional Problems in School-Age Children with ConHD

Children with somatic conditions, such as ConHD, are at risk for emotional and behavioral maladjustment [14]. Already in 1990, DeMaso et al. [15] reported impaired psychological functioning in children with cyanotic ConHD, compared to a group of children who spontaneously recovered from a mild ConHD. DeMaso et al. [15], however, used a rather global assessment instrument.

After 1990, the Child Behavior Checklist (CBCL) [16] has been used very often to assess emotional and behavioral problems in ConHD children [13].

The CBCL is a standardized parent report, covering a wide range of emotional and behavioral problems in children aged 6–18 years. It consists of eight specific subscales, two broad problem areas (internalizing and externalizing), and a total problem score. Internalizing consists of the subscales: anxiety/depression, withdrawn, and somatic complaints. Externalizing consists of the subscales: rule-breaking

(delinquent) and aggressive behaviors. The subscales social problems and thought and attention problems belong neither to the internalizing nor to the externalizing scale [16].

Multiple studies showed that parents of children with ConHD reported more total problems in their children compared to normative groups [17–23]. A meta-analysis from Karsdorp et al. [24] yielded a medium estimate of effect size for total problems in children with ConHD [24].

Several studies showed significant higher levels of internalizing problems in children with ConHD in comparison to normative samples, increased levels of withdrawn behavior [17, 25, 26], somatic complaints [17, 25–27], as well as anxiety and depression [17, 19].

Externalizing problems have also been reported for ConHD children [17, 20].

According to the meta-analysis of Karsdorp et al. [24], internalizing problems are found more often in children with ConHD than externalizing problems. Karsdorp et al. revealed a medium estimate of effect size for internalizing problems, but only a small estimate of effect size for externalizing problems [23].

Attention, social, and thought problems. Also important, studies using the CBCL also indicated elevated levels of attention problems [17, 22, 26], social problems [17, 25, 27], and thought problems [17, 25] for children with ConHD. Attention and social problems are clinically relevant since they may be associated with problems at school (see the paragraph in this chapter on intellectual and school functioning).

Reviewing these outcomes, it is important to consider that early studies included patients operated for ConHD in the 1970s, whereas most recent studies included patients operated after 1990. Since 1990, many aspects of medical treatment for ConHD had been improved. All these changes were believed to result in improved cardiological outcome. Therefore in Rotterdam, the Netherlands, a cohort study was started to test the hypothesis that improvements in medical care would also result in more favorable psychosocial outcomes for children with ConHD [28]. No study had thus far compared directly *long-term emotional and behavioral outcomes* of children treated more recently (operated after 1990) to those of *same-age patients*, operated before 1980 (“historical sample”). The recent and historical sample in the study consisted of the four same diagnostic groups: atrial septal defect, ventricular septal defect, transposition of the great arteries, and pulmonary stenosis [28].

The study revealed that the level of emotional and behavioral problems of the recent sample was comparable to that of the historical sample. Also frequency of contact with seeking support from mental health professionals for psychosocial problems of the child related to the ConHD was comparable in children operated before 1980 versus those operated after 1990. Especially internalizing problems, social problems, and attention problems were prevalent in both the more recent (>1990) and the historical (<1980) cohort. Surprisingly, in both cohorts, type of cardiac diagnosis had no influence on the level of behavioral and emotional problems [28].

Also noteworthy, in both cohorts, adolescents with ConHD reported more behavioral and emotional problems regarding themselves than their parents did about

them. Since ConHD children and adolescents tend to suffer more from internalizing problems (than externalizing problems), these problems are less visible to their parents, and these ConHD youngsters may tend to keep their problems to themselves (possibly not to burden their parents).

Summarizing, despite the major advances in medical treatment, no clear improvement on behavioral/emotional outcomes of the children was found. Unfortunately, children with ConHD who were treated more recently still seem to experience the same level of emotional and behavioral problems compared to children operated in former days [28].

3.3.2.2 Health-Related Quality of Life in School-Age Children with ConHD

In the last decade, interest for health-related quality of life (HRQoL) in children with ConHD has been growing. Sometimes, the terms health-related quality of life (HRQol) and health status have been used as though they were almost equivalents. Health status, however, refers to assessment by a person of his or her actual, more objective problems and limitations in functioning, whereas HRQol includes the person's subjective, emotional evaluation and reaction to such problems and limitations [7].

A number of studies have been done into HRQol in children with ConHD [29–31]. Studies often showed methodological problems, such as use of limited samples (e.g., only one diagnostic group involved), and variation in methods across studies. In general, it is known that children and parents may have different views about the impact of illness.

Spijkerboer et al. [7] performed a study in which HRQol was assessed both by children and parent ratings in a cohort of children with ConHD operated after 1990. In their study, they used the TNO-AZL Child Quality of Life Questionnaire (TACQOL). This instrument assesses general aspects of HRQoL in children aged 8–15 years [8]. An advantage of this instrument is that it does not only measure the occurrence of functional problems but also the *subjective evaluation* regarding those problems. Thus if a problem occurs, then negative emotional reactions are assessed, too.

Spijkerboer et al. found [7] that 8–11-year-old ConHD children reported significantly poorer HRQoL than reference peers on five domains: motor functioning, cognitive functioning, autonomy, social functioning, and positive emotional functioning. The parents reported poorer HRQoL regarding their children for cognitive functioning and positive emotional functioning compared to normative parents.

Furthermore, this study showed that children and adolescents with ConHD reported a poor HRQol regarding themselves than their parents did regarding them.

Moreover, cardiac diagnosis and type of invasive procedures (cardiac surgery versus catheter intervention) did not influence the level of HRQol of the children.

Summarizing, these findings indicate that 8–11-year-old ConHD children were at risk for poorer HRQol, both according to their parents as to their self-reports.

3.3.2.3 Intellectual and School Functioning in School-Age Children ConHD

These topics are mentioned here only very briefly, since this book contains a separate chapter on neuropsychological aspects in children with ConHD (by Miatton).

In another study of Spijkerboer et al. [32], it was found that, although the mean total IQ score was within the normal range, significantly poorer scores were found regarding verbal IQ and verbal comprehension for the total sample ($N = 117$) of ConHD children compared with the normative group.

In addition, 14 % of the total sample appeared to receive special education services, and 39 children (33 %) had repeated a school year in the past [32].

In previous studies, IQ was often assessed without measuring behavioral and emotional problems at school. In this study, no differences were found in teachers' reports of school-related behavioral and emotional functioning for ConHD children versus normative groups, apart from one exception: teacher reported more somatic complaints for ConHD children [32].

For clinical implications on intellectual and school functioning, see Sect. 3.2.5.

3.3.2.4 Motor Functioning in School-Age Children ConHD

Children and adolescents with ConHD may suffer from reduced exercise capacity and lower physical activity levels, possibly resulting in lower self-esteem and reduced health-related quality of life. Physical limitations may lead to social isolation and fewer possibilities to develop social competencies [33]. Recently Dulfer et al. wrote a review, investigating the association between exercise capacity, physical activity, and psychosocial functioning of ConHD youth [33].

They found that no clear relationships were found between exercise capacity, physical activity, and QoL in children and adolescents with ConHD.

E*xercise capacity* was strongly related to *physical* domains of QoL (both parent and self-reports of youngster with ConHD), but almost never to *psychosocial* domains of QoL. Physical activity as such was rarely associated with Qol, neither with the physical nor with psychosocial domains of QoL [33]. Remarkably, self-reported depressive symptoms were associated with both physical and psychosocial quality of life.

The few studies that have been executed into exercise training programs for ConHD youngsters showed promising results as to QoL and emotional and behavioral problems. However, the studies performed until now showed methodological flaws, so further research in this field is needed [33].

3.3.2.5 Clinical Implications, Preparing for Cardiac Procedures and Psychotherapy

Behavioral/Emotional Problems, HRQoL, Intellectual and School Functioning, and Motor Function

Reviewing the paragraphs above, we conclude that children with ConHD are at risk for behavioral/emotional problems, a reduced HRQoL, intellectual and school problems, and poorer motor function compared to healthy peers. On HRQoL domains,

ConHD children reported poorer positive emotional functioning; poorer cognitive, motor, and social functioning; and poorer autonomy compared to normative data.

What is the role of clinical psychologists in screening, diagnosing, and intervening regarding these problems?

In 2012, an extensive scientific statement from the American Heart Association was published regarding surveillance, screening, evaluation, and management strategies for neurodevelopmental and behavioral problems in children with ConHD [34]. We refer to this publication [34], since it provides a thorough and very detailed description, on which domains to screen, what (neuro)psychological instruments to use, and referral for psychological treatment, for children with ConHD with different diagnoses, from birth through adolescence. All psychological instruments recommended in that document have been used previously for evaluation in ConHD children and adolescents [34].

According to this statement, behavior should be monitored at every medical home visit from infancy through adolescence, by risk factor analysis, history gathering, and observation. In this statement, developmental screening based on age (e.g., 9, 18, 24, 30, and 48 months) is recommended for all children with ConHD. When concerns are detected in surveillance or during the developmental screening, behavioral screening can be useful [34].

Screening for both developmental and behavioral skills at the 30- and 48-month visits is especially relevant, since this may help in the early identification of the symptoms of learning and behavior disorders, well known to occur during school age in ConHD children (e.g., learning disabilities and attention-deficit/hyperactivity disorder: ADHD) [34].

In our opinion, it is very important to perform an early developmental and behavioral screening, since behavioral and emotional psychopathological problems are less persistent and better treatable at a younger age. By a standard follow-up screening, children at risk for neurodevelopmental and behavioral problems can be identified. Then adequate psychological service can be provided to prevent the development of additional problems.

Multi-informant approach. Furthermore we recommend a multi-informant approach, thus gathering information regarding psychosocial problems of the ConHD child from the child itself, the parents, and also the teacher. Especially inclusion of self-reports is important since previous studies [7, 35] (see Sects. 3.2.1 and 3.2.2) showed that children themselves reported more behavioral and emotional problems and problems on HRQoL domains than parents did about them.

Teacher reports [16] can be very useful to get insight into attention and learning problems of the child. Visuospatial problems [34] have been reported for ConHD, which may be associated with, for example, problems with writing and fine motor functioning. Clinical psychologists can act as a link between parents and school regarding learning problems of the child or discuss possibilities, for example, remedial teaching for the child.

As for *social functioning*, clinical experience learns that it is important to stimulate ConHD children to participate in social activities, practice social skills, and build friendships. For children with limited exercise capacity, who have difficulty

keeping up with peers in sports activities, other compensating activities can be very useful (e.g., making music, playing theater, chess, photograph or drawing/painting lessons, other creative activities, etc.). We would recommend compensating activities in a team of group format, since this facilitates making friends. To perform compensating activities is important for strengthening self-esteem and emotional resilience.

As for *motor functioning*, Takken et al. [36] have published recommendations regarding participation in leisure sports, physical activity, and exercise training programs for optimal physical, emotional, and psychosocial development for children and adolescents with ConHD.

Adolescents with ConHD do not achieve 60 min of recommended daily moderate-to-vigorous physical activity [37] and tend to become more obese.

Therefore, it is important to stimulate them to sport and participate in physical activities and to enhance their physical condition and overall feeling of well-being.

Monitored physical training or a cardiac rehabilitation program may help patients with poor physical condition to feel more secure about their bodily functioning and to discover their physical boundaries.

Preparing Children and Adolescents for Invasive Cardiac Procedures

In 2003, an extensive review was written on this topic [38] to which we refer. For different ages, useful interventions and tools are described in order to prepare children to invasive cardiac procedures, such as:

- Information giving: verbal/written/AV–video–CD-ROM preoperative classes hospital tours, *Internet resources*
- Cognitive behavioral techniques
- Biofeedback
- Play therapy
- Attention diversion/distraction
- Peer modeling

According to the review, better efficacy is gained by combining pre-procedural information with cognitive behavioral techniques. Children with previous medical experience are at higher risk for emotional problems.

Psychotherapy

In case of emotional behavioral disorders (such as anxiety disorders), cognitive behavioral therapy is the state-of-the-art treatment nowadays.

Cognitive behavioral therapy techniques involve, for example [38]:

- Progressive muscle relaxation
- Conscious breathing techniques
- Guided imagery
- Changing negative into positive thoughts; positive self-talk
- Exposure, step plans. Reinforcing positive behavior

During the last decade, for traumatic events resulting in posttraumatic stress symptoms or posttraumatic stress disorders, Eye Movement Desensitization and Reprocessing (EMDR) [39] became available. Empirical evidence on the efficacy of EMDR is scarce, though clinical psychologists have reported beneficial effects for traumatized children [39].

Through clinical experience it can be observed that psychotherapeutic treatment such as psychodynamic-oriented psychotherapy can be very useful to give insight in mechanisms of coping and adjusting to their disease for youngsters with ConHD.

3.3.3 Psychosocial Aspects for Adolescents and Transition-Related Topics

During the phase of adolescence, several key developmental experiences occur. Physical and sexual maturation, capacity for abstract reasoning, development of identity, striving for autonomy, social and economic independence, establishing (sexual) relationships, and moving towards adult roles are important challenges during adolescence. In this phase, it can be very difficult for parents to let go and to encourage autonomy in their child. Nonetheless, it is very important that parents stimulate their teenagers gradually to take on responsibility for their own health and to foster independence in their child. The adolescent's chronological age is not an accurate predictor of emotional maturity. Hospitalizations, physical limitations, and parental protectiveness may have hampered developmental tasks. In times of disease exacerbation, adolescents may show *regression*, and parents may return to their previous, more active role; this shift is normal [40].

Especially the *transitional period* from pediatric service to an adult department is a complicated process, for patients, parents, and pediatric and adult cardiologists. An educational goal of transitioning is to assist adolescents in achieving a basic understanding of their own condition [40]. Many adolescents have little understanding of their heart condition.

An adolescent health-care passport [41] may serve as a useful tool and as a lesson plan. The act of filling it out and keeping it up to date as patients gradually move through adolescence fosters competence and taking responsibility for their own care. Topics which should be discussed are diagnosis, procedure, medication, contraception, exercise, prophylaxis, medical follow-up, and insurance.

The British Cardiac Society Working Party [42] published a list of lifestyle problems on which specialist advice is often needed. The topics included are as follows:

- Contraception/marriage/pregnancy
- Genetic risks
- Noncardiac surgery/trauma
- Employment (fire or police service, army)
- Life and health insurance
- Driving license/pilot's license

- Psychosocial problems
- Issues with law/prison
- Hobbies
- Drugs, alcohol, smoking, obesity

Noncardiac primary health-care problems and advice concerning healthy lifestyles to prevent acquired heart disease should be discussed (drugs, alcohol, smoking, obesity) and are also noteworthy.

3.3.4 Clinical Implications: Adolescents and Transition-Related Topics

Limit testing and risk taking are normal hallmarks of adolescence. Adolescents want to feel in control of themselves, and noncompliance with medication regimen can be a result. It is recommended for health providers to be supportive and empathic, but also to be firm in explaining teens the consequences and discuss which aspects of their treatment are negotiable and which ones are nonnegotiable.

In 2012, the American Heart Association [34] stated that routine screening for psychosocial adjustment problems by primary care practitioners is likely to be adequate for the majority of adolescents with ConHD. Using a multi-informant approach (adolescents, parents, teachers) and multiple, well-established, psychometrically sound instruments provides a more comprehensive evaluation of the adolescent's psychosocial and mental health. In case of clinically significant problems, adolescents with ConHD should be referred to an appropriate behavioral or mental health specialist, preferably with expertise in the specific area of congenital heart disease.

In our opinion, it is very important that teams working with ConHD patients are multidisciplinary and able to offer specialized professional psychological and social counseling to these youngsters.

3.4 Clinical Case

To end this chapter we want to present a clinical case of a female adolescent, Joanna, referred to the Psychosocial Care Unit in our hospital, the Erasmus MC – Sophia Children's Hospital.

Rather than presenting a clear-cut success story, we end this chapter with a clinical case for which we decided:

1. Firstly, to intervene immediately with cognitive behavioral techniques
2. Thereafter, to perform extensive diagnostic psychological assessment

Since the editors of this book are Italian, we want to illustrate that, whereas normally a psychological intake starts with psychodiagnostics, many ways can lead to Rome.

Joanna, a 16-year-old girl with a medical history including premature birth (35 weeks of gestational age) and congenital heart defects (coarctation aortae and VSD), was referred to our unit. She underwent cardiac surgery twice (at 0 and 13 years of age).

During a recent outpatient follow-up examination, Joanna tells her pediatric cardiologist that she suffers from panic attacks and (symptoms of) hyperventilation. This induces very stressful anxious thoughts that something is wrong with her heart. In addition, she fears that she has to undergo a third cardiac surgery in the near future. Since the panic attacks and hyperventilation complaints cannot be understood from a cardiac point of view, Joanna is referred to a child psychologist for psychodiagnostic assessment and psychological treatment.

After 2 weeks, Joanna and her mother come to the child psychologist for the first intake session. Joanna tells about her anxieties and worries as to her heart condition. She explains how and what she thinks and feels (physically and emotionally) when experiencing panic and hyperventilation attacks. Anxious (nonrealistic) fantasies are reported, including that the stitches put in her heart during the last cardiac surgery might burst open. She also believes she can feel her blood flow through her heart.

Furthermore, Joanna feels ashamed and often embarrassed by the cardiac scars on her chest (resulting in not wanting to wear a bikini). Joanna often does not feel understood by family members or friends. She does not want to bother or worry other people when feeling sad and down.

Her mother mentions Joanna is easily irritated, emotionally moved (crying), and unhappy.

Every once in a while, she suffers from nightmares. At school Joanna has problems with her physical education (PE) teacher. She suffers from the fact that this teacher gives her negative feedback and poor grades, despite the fact that she really tries to do what she can during the gymnastic lessons. Since Joanna has insufficient exercise capacity, she receives treatment and training from a physical therapist to improve this. Although she benefits from this exercise training, which has a positive effect on her fitness and exercise capacity, the panic attacks and hyperventilation are still present and negatively influencing quality of life.

In the first phase of psychological treatment, we decide to intervene directly on the hyperventilation and panic attacks with cognitive behavioral techniques, since this was the most urgent need. Extensive psychodiagnostic assessment was planned as the second step.

Regarding the panic attacks, psychoeducation made Joanna gain insight, understanding, and belief that the panic attacks were psychological in nature and not an indication of a malfunctioning heart. Furthermore, relaxation techniques and cognitive restructuring were taught. Cognitive restructuring included: monitoring, evaluating and detecting negative thoughts and replacing them by positive ones. These techniques resulted in a decrease of the panic attacks. Joanna was happy to feel less insecure and to be more in control regarding the panic attacks. Gradually, it appeared and seemed that disease-specific (cardiac-related) anxieties were not the only problem, but that generalized anxiety might also be present. At this point we decided, in

consensus with Joanna and her mother, to perform extensive further psychodiagnostic assessment, which is now being executed.

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Adjusting to Congenital Heart Disease in Adolescence: The Role of Patients' Personality and Self

4

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During the last couple of decades, major advances in medicine have resulted in an increasing number of adolescents and emerging adults living with congenital heart disease (ConHD) [1]. The emergence of this growing population poses new challenges to healthcare because many of these youngsters suffer from the medical and psychosocial consequences of their illness. Indeed, *adolescents* with ConHD are generally seen as a high-risk group in terms of psychosocial functioning [2]. In addition to several physical and neurocognitive changes, role expectations change substantially during this period in life. Adolescents are expected to form intimate friendships, to grow independent from parents and establish a social network of their own, and to develop into mature individuals adopting various social and societal roles [3]. These multiple changes in biological and psychosocial domains are generally found to be reflected by changes in *personality* and *self-concept*, as adolescents tend to move toward a more mature sense of self [4]. However, adolescents with ConHD not only have to deal with these developmental demands; they are confronted with several illness-specific challenges as well. For instance, many patients struggle to cope with the uncertainty regarding illness course and prognosis, difficulties with fitting into the peer group, symptom burden (e.g., cyanosis, lack of energy, and shortness of breath), and physical activity restrictions [5]. Hence, studies are increasingly focusing on patients' *quality of life* and *self-rated health*. Self-rated health has been shown to be a powerful predictor of health

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outcomes – including mortality – in both community and patient samples, even after accounting for objective health status, health behaviors, and sociodemographic factors [6].

To understand why some patients show poor quality of life and self-rated health whereas others display signs of resilience, research on potential determinants of quality of life and self-rated health is urgently needed. A potentially important determinant that has not received much attention to date is patients' personality and self-concept. Extensive research has linked Type A (characterized by hostility, time urgency, and competitiveness) and Type D personality (characterized by negative affect and social inhibition) to mortality and adverse health outcomes in patient groups with acquired cardiovascular pathologies [7–9]. In contrast, the personality and self-concept of individuals with ConHD has received little to no attention in the literature. This chapter therefore aims to summarize the current research literature on personality and self-concept in adolescents with ConHD.

4.1 A Brief Introduction into Personality and Identity

One of the most important developmental tasks during adolescence is establishing a strong and mature sense of self [3]. According to a recent model of personality development, an individual's sense of self has to be assessed at different levels of analysis to obtain a thorough view on his or her personality configuration [10]. *Personality traits* are typically seen as constituting the basic level of analysis. These traits describe the most fundamental personality differences between individuals that account for consistencies in how they act across situations and over time. However, people do more than merely act in more-or-less consistent ways as determined by these relatively enduring or stable personality traits. As agents of their own development, people make motivated identity choices, plan their lives, and strive for certain goals [10]. For instance, although an individual may be characterized by a lack of self-discipline in various areas in life (and, hence, might score relatively low on conscientiousness), he or she might be motivated to achieve certain goals in a specific area in life which he or she deems especially important for his or her personal identity. Hence, it is critical that studies focus on both personality traits and other self-related variables such as *identity*, representing important core (or relatively stable) and surface (or more malleable) characteristics of an individual's sense of self.

4.1.1 Personality Traits as Core Characteristics of the Self

Nowadays, most researchers agree that the basic level of personality can be subsumed under five broad traits: extraversion (energy, sociability, and experiencing frequent positive moods), agreeableness (kindness, empathy, and cooperativeness), conscientiousness (self-discipline, organization, and responsibility), emotional stability (the ability to deal with negative emotions), and openness to experience (the way an individual seeks and deals with new information) [11].

These *Big Five* personality traits have been proven valuable predictors of physical and psychological health in both community and patient samples [12–15]. In adolescents with asthma, for instance, several of the Big Five personality traits were found to predict patients' quality of life above and beyond the effects of sex, age, ethnicity, and education level [14]. In youngsters with type 1 diabetes, similar findings were obtained. That is, the Big Five personality traits were found to predict several problem areas in diabetes above and beyond the effects of sex, age, and illness duration [15].

Personality traits are believed to predict these health outcomes in both a direct and indirect manner. According to the psychophysiological model, for instance, the tendency to experience stress and negative affectivity may directly affect an individual's metabolism, immunity, and cardiovascular system [16]. However, personality traits may also predict these health outcomes indirectly through their relation with coping, illness perceptions, health-threatening behaviors (such as substance abuse), and treatment adherence (in case of chronic illness) [16]. Adolescents low in emotional stability and conscientiousness, for instance, have been found to report higher rates of noncompliance with treatment guidelines, putting them at risk for future health complications [17]. Such knowledge allows healthcare professionals to intervene before patients become nonadherent and engage in health-threatening behaviors. Furthermore, individuals low in emotional stability and conscientiousness have been found to use more avoidant and passive ways of coping in dealing with daily stressors, including illness-specific challenges [15]. These passive and avoidant ways of coping, in turn, have been linked to illness-related problems [15].

4.1.2 Identity Dimensions and Statuses as Surface Characteristics of the Self

Besides the Big Five personality traits constituting a first or basic layer of the self, processes of personal identity formation constitute an additional layer of the self, especially in adolescence [10]. Adolescents have to address the self-defining question: "Who am I and where do I want to go with my life?" Identity formation is typically measured using the key dimensions of *exploration* and *commitment* [18]. Whereas exploration entails a search into different life alternatives (e.g., "Do I want to become a psychologist or a lawyer?"), commitment signifies the adherence to a specific choice and the implementation of this choice in daily life.

Using these different dimensions, recent studies have identified five to six *identity statuses* (reflecting different ways an individual can address identity-related questions at a certain point in time) in both community and patient samples [19, 20]. Individuals in the *achievement* status typically showed firm identity commitments preceded by a thorough exploration of identity alternatives. For instance, they want to become lawyers because they thoroughly reflected about it and fully identified with this choice. As such, they achieved an identity through a period of experimentation and exploration. Similar to individuals in the achievement status, individuals in the *foreclosure* status showed strong identity commitments. However,

these commitments were made without much prior exploration. For instance, they want to become lawyers simply because their mother or father is a lawyer. Although individuals in the achievement and foreclosure statuses seem to function quite similarly on variables such as self-esteem, achieved individuals who explored various possibilities and chose the one option that suited them best typically score higher on variables such as life satisfaction and intrinsic motivation [21]. Hence, although having strong identity commitments benefits individuals in their daily lives, having commitments that are self-endorsed (as is the case especially in the achievement status) leads to even better functioning in certain life areas.

The remaining statuses consisted of individuals characterized by relatively weak identity commitments. In contrast to the achieved and foreclosed statuses, these individuals lacked strong guiding commitments in daily life. Individuals in the *moratorium* status were currently uncommitted but they launched themselves in the exploration process to seek out various identity alternatives. This exploration, however, was often accompanied by high distress and worries over the future. Finally, individuals in the diffusion statuses also scored low on commitment but, as opposed to the moratorium individuals, they did not engage themselves in a purposeful exploration process. Individuals in the *carefree diffusion* cluster rather seem to enjoy this uncommitted state, live from day to day, and do not feel the urge to engage in a lot of identity work. Individuals in the *troubled diffusion* cluster, on the other hand, continuously worry where their lives would lead them, but, due to a lack of both internal and external resources, they do not succeed in proactively tackling the identity questions they are facing. They seem to be stuck momentarily in a state of worry and rumination.

Previous research in community samples has demonstrated that the ways in which adolescents tackle identity issues can have important implications for their daily functioning [19]. More specifically, adolescents who kept on postponing identity-related decisions or kept on worrying about where their lives should lead them were found to experience various psychosocial difficulties, such as increased levels of depressive symptoms and lowered self-esteem. In contrast, adolescents who purposefully explored various future possibilities and succeeded in committing to certain life decisions were found to be the most resilient individuals. Yet, few studies to date have looked at associations between identity processes and illness functioning in adolescents with chronic illness. In one of our studies in emerging adults with type 1 diabetes, identity processes were differentially related to the ways in which patients coped with and adjusted to their illness [20]. More specifically, the identity statuses representing a strong sense of identity were accompanied by fewer diabetes-related problems and depressive symptoms and more adequate coping strategies.

Taken together, the findings obtained from previous research in community and patient samples indicate that the establishment of a strong personalized identity and the development toward a mature personality profile are closely related to adolescents' physical and psychological health. Hence, we believe that research looking at the physical and psychological health of adolescents with ConHD should pay more attention to these key developmental tasks.

4.2 What About Individuals with Congenital Heart Disease?

Despite the fact that extensive research has linked *Type D personality* (which is basically a combination of low extraversion and low emotional stability) to mortality and adverse health outcomes in patient groups with acquired cardiovascular pathologies [8, 9], the personality and self-concept of adolescents with ConHD has received little to no attention in the literature. For instance, few studies to date have examined in depth whether there are systematic differences between patients and controls in terms of their personality. However, it is crucial for future research to examine whether these patients have an increased risk for developing certain *self-related vulnerabilities*, given that a strong sense of self has been repeatedly associated with patients' physical and psychological health [22, 23].

According to a recently forwarded model of *person-environment transactions*, changes in an individual's personality and self-concept can be triggered by changing roles, life events, and/or daily challenges [24, 25]. For instance, making the transition from high school to college can lead to increases in independent functioning. Along these lines, having a chronic illness such as ConHD can be expected to shape youngsters' sense of self, given that chronic illness is typically conceptualized as a *biographical disruption* requiring a fundamental rethinking of an individual's self-concept [26]. For instance, adolescents with ConHD (and especially those with a more complex defect) might report lower levels of emotional stability, due to the fact that they have to deal with the additional challenges imposed by their illness. In the next sections, we review the limited research focusing on comparisons between individuals with and without ConHD in terms of personality and identity.

4.2.1 Differences in Personality and Identity Between Patients and Controls

Personality. A recent study by Schoormans and colleagues [27] was the first to date to investigate in depth the personality of individuals with ConHD. More specifically, this study examined the *point prevalence* of Type D personality (defined as a cutoff score of 10 or more on the social inhibition and negative affectivity subscale of the DS14 and, thus, operationalized as a dichotomous variable) in a large sample of adults with ConHD. Approximately 20 % of patients were found to score above the cutoff of having a Type D personality, which is quite similar to the prevalence of Type D personality in the general adult Dutch population (18–21 %) [28]. Interestingly, this point prevalence is slightly lower than the one found in other cardiac populations, such as adults with myocardial infarction (24 %) or congestive heart failure (25 %) [29, 30]. The slightly higher prevalence of Type D personality in cardiovascular patients might stem from the fact that Type D personality has been linked to the pathogenesis of acquired heart diseases [8, 9]. Furthermore, Schoormans and colleagues [27] did not observe any differences between Type D and non-Type D patients in terms of illness complexity, as conceptualized by Task Force 1 of the 32nd Bethesda conference [31]. Hence, patients with a more complex

defect were not more likely to display a Type D personality profile as compared to patients with a defect of simple or moderate complexity. Although this recent study has provided us with important insights, personality research in cardiac disorders should be expanded beyond assessing Type D personality.

As mentioned earlier, an alternative framework that can be valuable for research aimed at understanding linkages between personality and illness functioning is the Big Five of personality. In a recent study at our center [22], we explored similarities and differences in Big Five scores between a large sample of adolescents with ConHD and a sample of healthy adolescents matched (1:1) on sex and age. This study was part of a larger project called *i-DETACH* (information technology Devices and Education program for Transitioning Adolescents with Congenital Heart disease), a project in which adolescents between the age of 14 and 18 are followed over a period of 4 years. Each year, these adolescents are asked to fill out questionnaires concerning their physical and psychosocial health, personality characteristics, and identity processes. The findings of this study suggested normalcy rather than deviance in patients' personality profiles, as few differences were found in Big Five scores between adolescents with and without ConHD. Nonetheless, adolescents with ConHD were found to score significantly lower on *extraversion* as compared to healthy controls. As activity level constitutes a central feature of extraversion [32], patients' lower levels of extraversion might be partially explained by their lower *activity level*, resulting from the physical complaints accompanying their illness (e.g., dyspnea). Similarly, *positive affectivity* is generally considered one of the central features of extraversion [32]. Possibly, adolescents with ConHD experience fewer positive emotions, having to deal with the worries and challenges of their illness, further lowering their extraversion scores. However, little systematic knowledge is available on the extent to which adolescents with chronic illness (and with ConHD in specific) experience positive affect, so this latter explanation needs to be addressed in future research efforts. A final alternative explanation for patients' lower extraversion scores could be found in the process of *normalization*, which is commonly observed in adolescents with ConHD. Normalization refers to patients' struggle with themselves and their environment to be accepted as "normal" [33]. From this point of view, an extraverted personality might be less warranted for adolescents with ConHD who do not want to stand out and just want to be like everybody else. Indeed, Rigby and Huebner [34] already pointed out that the desire not to stand out might reduce the advantages of high extraversion among certain adolescents.

Finally, although one might expect adolescents with a heart defect of greater complexity to face additional stressors that may impact upon their personality development, no mean differences in Big Five scores were observed between adolescents with a simple, moderate, and complex heart defect. Hence, no evidence was found for delayed personality maturation in adolescents with a more complex heart defect.

Identity. In two of our recent studies [23, 35] based on the i-DETACH project, similarities and differences in identity formation were examined between adolescents with ConHD and a sample of healthy adolescents matched (1:1) on sex and age. Patients were found to show lower levels of both *identity exploration*

and *worry* as compared to their healthy peers. More specifically, adolescents with ConHD were slightly underrepresented in the moratorium and troubled diffusion statuses and slightly overrepresented in the carefree diffusion status. Possibly, some of these patients perceived fewer opportunities to explore identity-related issues when dealing with the challenges of their illness. Furthermore, patients might be less inclined to reflect on future options because of *perceived restrictions on future possibilities* due to their illness [36]. However, despite these minor differences, adolescents with ConHD were found to tackle identity issues in a way similar to what their peers did, testifying to the *resilience* these patients display. Furthermore, similar to the findings presented above on the Big Five personality traits, the different identity statuses were unrelated to the complexity of patients' heart defect. Hence, more complex diagnoses do not seem to be associated with a more problematic identity formation process.

Taken together, these findings show that, although growing up with ConHD constitutes a big challenge for some adolescents, patients are generally as competent as their peers in addressing the developmental task of identity formation and in developing a mature sense of self. Nonetheless, some patients were found to struggle with establishing a strong sense of self, which then might negatively impact on their physical and psychological health. Indeed, although personality and identity are not implicated in the pathogenesis of ConHD, they could potentially affect patients' prognosis, health outcomes, and illness functioning. That is, mature personality characteristics (e.g., being conscientious and emotionally stable) could help patients in dealing with illness-related challenges, thereby playing favorably into their illness functioning [14]. Conversely, certain personality characteristics (e.g., the tendency to experience negative affect) may put patients at risk for illness-related problems such as the inability to deal adequately with certain life restraints due to their condition. In the next sections, we review the limited research focusing on the role of personality and identity for patients' physical and psychological health.

4.2.2 The Role of Patients' Self for Their Physical and Psychological Health

Personality. In the aforementioned study by Schoormans and colleagues [27], patients with *Type D personality* were found to report poorer functional status, health status, and quality of life as compared to non-Type D patients. In addition, Type D patients tended to show less healthcare utilization, even after controlling for confounding variables such as illness complexity, functional status, health status, and quality of life. This failure to consult for cardiac symptoms is paradoxical, given that individuals with Type D personality have been found to worry more about their health status [37]. High levels of *social inhibition* – a key characteristic of Type D personality – might keep these patients from disclosing their worries to healthcare professionals [38]. In addition, individuals with Type D personality tend to use more passive and avoidant ways of coping, which makes them less likely to act

upon the symptoms they are experiencing [39, 40]. Previous research has suggested that such an inadequate consultation behavior might be a mechanism underlying the negative relationship between Type D personality and health outcomes [37].

In one of our own studies [22], which was again based on the i-DETACH project, important associations were uncovered between the Big Five personality traits, quality of life, and self-rated health in adolescents with ConHD, even after controlling for the effects of sex, age, and illness complexity. *Emotional stability* turned out to be the strongest predictor of quality of life and both generic and illness-specific domains of self-rated health. This finding can be partially explained by the tendency of individuals low in emotional stability to experience negative affect [41] and to focus on and report more symptoms [14, 42]. Additionally, these persons tend to use dysfunctional coping when confronted with stress [14], making them more vulnerable for poor illness functioning. Next, also *agreeableness* and *extraversion* were found to be unique predictors of patients' quality of life and self-rated health. Individuals high in agreeableness and extraversion are generally found to show adequate coping skills, that is, they tend to hide their illness less for others, use more positive reappraisal, and/or show less activity avoidance [6]. Moreover, these individuals typically receive more social support [41, 43], which has been shown to be an important determinant of quality of life in individuals with ConHD [44].

Conscientiousness appeared to be a unique predictor of patients' emotional, cognitive, and school-related functioning. Interestingly, however, conscientiousness did not predict patients' quality of life. Extensive research has found conscientiousness to affect individuals' health and adjustment through relationships with both harmful and protective health behaviors, including self-management in individuals with chronic illness [45, 46]. Although self-management may help adolescents with ConHD to prevent the occurrence of cardiac complications and coronary heart disease later in life [47], self-management is less central for obtaining favorable outcomes in the short run as compared to other chronic illnesses in which daily self-management is particularly critical (e.g., type 1 diabetes). Indeed, the majority of patients with simple or moderate heart defects are asymptomatic with no exercise or lifestyle limitations [6]. As a result, the quite limited predictive value of conscientiousness for patients' quality of life may be partially explained by the less central role of self-management in most ConHD patients. Finally, although *openness* was found to relate to quality of life and some domains of self-rated health, none of these associations remained significant after controlling for the other Big Five traits. Hence, the trait of openness seems to be less important for patients' illness functioning.

In sum, these findings demonstrate that the *Big Five* is a valuable framework for examining linkages between personality and health in patients with ConHD. Whereas illness-specific domains of self-rated health were mainly related to emotional stability, several personality traits contributed to patients' quality of life and generic domains of self-rated health. Therefore, future research in adolescents with chronic illness should not exclusively rely on rather narrow personality perspectives, such as Type D personality, as the present study has shown that also agreeableness and conscientiousness can be considered important predictors of patients' illness functioning [48].

Identity. Although adolescents were found to tackle identity issues in a way similar to what their peers did (as mentioned above), this general finding did not preclude some patients from being at risk for a problematic identity formation process. Over 10 % of the sample belonged to the *troubled diffusion* status, being the most problematic status characterized by a lack of steady identity commitments and increased levels of worry about the future [23]. These patients showed the highest levels of distress and the lowest quality of life and self-rated health. In comparison to *achieved* and *foreclosed* patients – both characterized by strong identity commitments and few worries over the future – patients in this *troubled diffusion* status scored higher on depressive symptoms, loneliness, and emotional and cognitive problems and experienced more academic difficulties. Furthermore, not only did these adolescents score worse on these relatively generic outcome measures, they also experienced the highest levels of treatment anxiety and reported the most problems in communicating with clinicians, suggesting that adolescents in the troubled diffusion status may be particularly at risk of being *lost to follow-up*. These latter findings are very important given that 21–76 % of patients are lost to follow-up by the time they reach adulthood and, hence, do not receive proper lifetime care, which is associated with significant morbidity [49, 50].

4.3 What Does All of This Mean for Clinical Practice?

Provided that continued research efforts also identify personality and self-concept as important determinants of physical and psychological health in individuals with ConHD, these findings can have important practical implications. First of all, individualized intervention programs can be developed which take into account the unique strengths and weaknesses of each patient. Recently, quite a lot of interest has arisen in these so-called *personality-informed interventions* or *personalized medicine approaches* [51]. By assessing the personality of patients, healthcare professionals are not only provided with a context for understanding the problems patients report, it can also help them to approach patients in a manner that fits their specific personality characteristics [52]. In addition, having a view on the core dispositions of the patient places healthcare professionals in a much better position to select appropriate interventions and to frame these interventions to the patient [52, 53]. Individual patient-level tailoring has not only been found to render interventions more effective, it may result in greater cost effectiveness as well [51].

A next step would be to improve patients' physical and psychological health directly through the modification of patients' personality traits and self-concept. Although personality traits have generally been conceptualized as stable and relatively unchangeable patterns of thoughts, feelings, and actions [11], emerging research has demonstrated that personality traits can be changed through interventions [54]. According to a recently developed framework [55], personality traits can be modified by targeting the core behaviors that underlie these traits (e.g., stimulating goal setting and self-discipline in individuals low in conscientiousness). Through repeated practice of new target behaviors, behavioral changes

may become more automatic and ingrained over time, ultimately manifesting themselves in trait-level changes. These trait-level changes, in turn, may affect patients' physical and psychological health. However, before implementing such interventions in clinical practice, more research is required on the appropriate dose (i.e., number of sessions) and format (i.e., group vs. individual) needed to change patients' personality traits [55].

Finally, the current findings could have important implications for *transition programs* aimed at assisting adolescents in transferring from pediatric cardiology to adult healthcare. The main goal of these transition programs is to maximize patients' functioning and well-being through high-quality, developmentally appropriate healthcare services [56]. A recent literature review emphasized that up till now no consensus has been reached on how to measure patients' *readiness for transfer* [57]. According to Moons and colleagues [56], an understanding of how well patients tackle normative developmental tasks (such as building a mature sense of self) can help healthcare professionals in determining the transitional needs of these patients and in developing the most appropriate and patient-oriented care. Hence, when deciding if and how to make the transfer, healthcare professionals may want to take into account the extent to which patients have set personal goals that are well-thought through and self-endorsed (indicative of a strong personalized identity), as well as the extent to which patients' behaviors are characterized by high responsibility, cooperation, sociability, and emotional stability (indicative of a mature personality profile).

4.4 How to Move the Field Forward?

Despite the fact that recent studies have substantially advanced our knowledge base on patients' personality and self-concept, there is still a lot of work ahead of us. First, little is known about the precise mechanisms through which patients' personality and self-concept could affect their physical and psychological health. Two potentially important intervening or mediating factors are patients' *illness perceptions* (i.e., how they think about their illness) and *coping strategies* (i.e., how they deal with illness-related challenges). According to the "Common Sense Model" (CSM), patients create their own models or representations of their illness in order to make sense of their illness which, in turn, influence their coping strategies and health outcomes [58]. Previous research in other patient samples has shown that coping and illness perceptions function as important mediating factors in the associations between personality and identity on the one hand and patients' physical and psychological on the other hand [14, 15, 20, 39, 46]. The study by Skinner and colleagues [46], for instance, provided evidence for the mediating role of illness perceptions in the relationship between personality and self-care in youngsters with diabetes. More specifically, they found that youngsters low in emotional stability typically believed diabetes to represent a serious threat to one's health which, in turn, was related to better self-care. Furthermore, youngsters high in conscientiousness typically believed that the treatment regimen would be effective in preventing

diabetes complications and in maintaining diabetes control, again resulting in better self-care. Although a very recent study demonstrated the predictive value of patients' illness perceptions for their quality of life 2 years later [59], few studies to date have focused on the illness perceptions and coping strategies of individuals with ConHD.

Third, most of the studies described in this chapter are cross-sectional in nature. *Longitudinal* studies in which adolescents with ConHD are followed over longer periods in time are needed to investigate the directionality of effects. For instance, it is typically assumed that adolescents who tackle identity issues in a maladaptive manner are at increased risk for illness-related problems. However, in line with the *scar model* (which states that psychosocial difficulties may lead to changes, or the so-called scars, in an individual's personality and self-concept), illness-related problems may also negatively impact on patients' identity formation process [60]. Indeed, previous research in community samples has shown these associations to be *reciprocal* in nature [19]. The aforementioned i-DETACH project might help us in answering these kinds of questions given the longitudinal four-wave design. Such longitudinal research would also allow us to chart the developmental trajectories of identity and illness functioning across time and to investigate how such trajectories influence one another.

Finally, the majority of studies focused between the patients' sense of self and their self-rated physical and psychological health. Future research may also want to look at associations with other potentially important outcome measures such as patients' *health behaviors*, *illness knowledge*, and *actual functional status*, all constructs with specific relevance toward patients with ConHD.

4.5 General Conclusions

In this chapter, two key issues were addressed. First of all, we examined whether adolescents with ConHD have an increased risk for developing certain self-related vulnerabilities due to the many additional stressors they are facing. Although some minor differences were observed between adolescents with and without ConHD (i.e., patients had lower extraversion scores and explored future possibilities less broadly as compared to healthy controls), patients were generally as competent as their peers in addressing the developmental task of identity formation and in establishing a mature sense of self. Hence, it was not the case that growing up with a chronic illness such as ConHD negatively impacted on the personality and self-concept of patients.

Second, we explored the role of patients' personality and self-concept for their physical and psychological health. Although extensive research has linked Type A and Type D personality to mortality and adverse health outcomes in patient groups with acquired cardiovascular pathologies, the personality and self-concept of individuals with ConHD have received little to no attention in the literature. In this chapter, the findings of three recent studies were discussed in which patients' personality and identity status were found to predict a variety of outcomes including

quality of life, self-rated health, and healthcare utilization, even after controlling for the effects of sex, age, and illness complexity. Hence, these findings suggest that patients' personality and self-concept might be important targets for future intervention and prevention efforts aimed at improving these patients' physical and psychological health. However, more research is needed before such interventions can be implemented into clinical practice. Nonetheless, we hope that the present findings have encouraged researchers to further study self-related variables in youngsters with ConHD and have sensitized healthcare professionals to take such variables into account when working with these adolescents.

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Neuropsychological Aspects in Children and Adolescents with ConHD

5

Marijke Miatton

5.1 Introduction

Advances in pediatric cardiothoracic surgical techniques have led to a significant decrease in mortality over the last decades, shifting the research focus within pediatric cardiology to morbidity [9]. Although a normal life expectancy is assumed in patients with early normalization of their cardiopulmonary status, negative developmental outcome might not supervene until later in life with an important impact on educational achievement, employment, relationships, and quality of life. Because cognitive advancements can be made by intervention provided in the first years of life [1], identifying *developmental deficits* and subsequent remedial interventions is of utmost importance in the *congenital heart disease* (*ConHD*) *population*. In this chapter, we will focus on the neurodevelopmental and cognitive functioning of children and adolescents with a ConHD. The chapter will be divided in three age groups: <3 years (early development), 3–6 years (preschool age), and 6–18 years (school age and adolescence). The neurodevelopmental and neuropsychological results will be described along different cognitive domains such as mental development, intelligence, attention, memory, language, visual perceptual and spatial skills, psychomotor skills, and executive functioning. Studies on children diagnosed with a syndrome known to affect motor or cognitive functioning such as trisomy 21 and the 22q deletion spectrum, studies on children with associated extracardiac anomalies, the need for heart transplantation, and low birth weight (<2.5 kg) are excluded. Further, a review of variables influencing the child's developmental abilities is not within the scope of this chapter. However, the change in

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search for *causal factors* from operative variables and intraoperative management to more patient-specific factors will be briefly outlined. In the 1990s and early 2000, the effect of deep hypothermic circulatory arrest, cooling time, hematocrit value, length of postoperative stay, and clinical seizures on neurodevelopment was thoroughly investigated. In many centers, the outcome of these studies led to adjustments in the surgical management of ConHD. Recently however, research proves that patient-specific and family-specific factors such as birth weight, ethnicity, presence of a genetic syndrome, parental style, etc., also substantially influence neurodevelopmental outcome [2, 3].

While infants undergoing more complex surgical procedures usually display poorer developmental outcome, merely assuming patients with an acyanotic ConHD to have a more favorable cognitive outcome remains a controversy. The idea of these children being free of neuropsychological deficits is challenged in several studies [4, 5] emphasizing the need for screening in all children with ConHD, including those with mild cardiac symptoms and seemingly normal neurodevelopment.

5.2 Early Development (<3 Years)

The *Bayley Scales of Infant and Toddler Development* is "the most commonly applied measurement tool for assessing early development" in both clinical and research settings [6]. The Bayley Scales monitor development and assess cognitive and motor outcomes. These scales display good psychometric properties [7] and are also commonly used in the ConHD population. *Mental development* of patients aged 1–3 years, undergoing cardiac surgery within the first 6 months of their lives, usually falls within one standard deviation of the test mean, reflecting a lower yet normal score. However, parental reports on children with a transposition of the great arteries (TGA) indicate poor expressive language at 2.5 years of age [8]. As far as *motor functioning* is concerned, scores usually fall between one and two standard deviations from the test mean, indicating clear deficits on this level [9, 10], especially for gross motor functioning [11]. Long et al. [12] point out that the new third version of the Bayley Scales (BSID-III) may underestimate developmental delay in 2-year-olds as more than 80 % of the infants scored within the normal range on each of the three composite scores (cognitive, language, motor). Using the Griffiths developmental scale and the Alberta Infant Motor Scale (AIMS), Sarajuuri et al. [13] found children aged 1 year with a univentricular heart to have a significantly lower level of motor development, while children with a hypoplastic left heart syndrome (HLHS) displayed a more widespread developmental delay. At 30-month follow-up, the same authors administered the BSID-II in these infants and reported a significantly lower *Mental and Psychomotor Developmental Index* (PDI) in HLHS patients compared to healthy controls. Patients with univentricular hearts only scored lower scores on the PDI [14]. Although performances on group level generally fall within normal expectations, a large variability within the sample exists. Infants undergoing more complex surgical procedures usually display poorer developmental outcome.

Accumulating evidence emphasizes the potential for an *antenatal or early postnatal onset* of neurological dysfunction. Prior to surgery, neurological abnormalities such as hypotonia, hypertonia, microcephaly, poor state regulation, lethargy, restlessness, jitteriness, and motor asymmetry have been identified [15]. In utero events impact early neurodevelopment and influence long-term outcome. Pre- and postoperative studies in children with ConHD indicate that neurological abnormalities prior to surgery predict adverse long-term outcome [16].

The *predictive value* of neurodevelopmental data in view of later school performance may be limited, due to different factors such as the lack of longitudinal and long-term follow-up studies, limited sensitivity of the scales used, individual variability in early skill development, and the fact that deficits might not supervene until later in life. However, several successful attempts have been made to clarify the relationship between early and late neurodevelopmental outcome and will be cited throughout this chapter.

5.3 Preschool Period (3–6 Years)

From the age of 4 years, more valid and standardized testing is possible. Most commonly used intellectual and neuropsychological measures in international research are the Wechsler Preschool and Primary Scale of Intelligence (*WPPSI*), the Beery Visual Motor Integration Test, the Peabody Picture Vocabulary test, the Movement Assessment Battery for Children, and the NEPSY (A Developmental Neuropsychological Assessment).

A thorough longitudinal follow-up study of infants who underwent an arterial switch operation for transposition of the great arteries with or without a ventricular septal defect is the *Boston Circulatory Arrest Study*. Follow-up data from this study comparing different surgical techniques (*circulatory arrest versus low-flow bypass*) revealed at the age of 4 years a continuation of the deficits in gross and fine motor functioning and in speech functions already identified at the ages of 1 year and 2.5 years. The group receiving circulatory arrest showed more oromotor and facial movement abnormalities, immaturity in hand use and gait, and abnormalities in speech production. The total group (circulatory arrest group and low-flow bypass group) performed significantly below population norms on intelligence, expressive language, visual-motor integration, motor planning and organization, and oromotor control [17]. At 5 years of age, *full-scale intelligent quotient* (FSIQ) after arterial switch operation is within the low-average normal limits [18], as is confirmed for most common ConHD such as TOF, ventricular septal defect, HLHS, and double outlet right ventricle [19–21]. Receptive language skills were also age appropriate in this study [20]. Mostly, patients' performances are compared with age- and sex-matched healthy controls. The patients usually score significantly lower than these controls, but it should be considered that overall their scores fall within the normal limits. Children with univentricular hearts, especially those with HLHS, are at higher risk for adverse outcome, with about 26–29 % of them presenting with major neurodevelopmental outcome [21].

Children with a cyanotic heart defect who underwent open heart surgery also appear to be at great risk for problems in the sensorimotor domain and verbal reasoning and to a lesser extent for visuospatial skills, number skills, and nonverbal reasoning [3].

Performances at the age of 2 years correlate well with performances at the age of 5 years in children with various ConHD. Early childhood mental scores lower than 55 are *predictive* for adverse mental outcome and intellectual impairment. Scores between 70 and 84 are often accompanied with learning difficulties. A mental development index (BSID-II) lower than 85 at 2 years correctly predicted a WPPSI FSIQ lower than 85 at 5 years of age in 85.7 % of the children [18]. These findings stress the need for early neurodevelopmental screening and validate their importance.

5.4 School-Age Period (6–18 Years)

Children with severe ConHD are confronted with multiple confounding risk factors such as poor cerebral perfusion, perinatal instability, more frequent need for surgery, and seizures, all negatively influencing cognitive functioning. A clear effect of disease severity on cognitive functioning is established, and cognitive functioning remains fairly stable across age groups [22].

5.4.1 Intelligence

Initially, investigators focused on the *intellectual consequences* of pediatric cardiac surgery. A first study on 38 children with various ConHD, assessed 22 months to 6 years after open heart surgery, reported normal intellectual capacities [23]. These results were confirmed in many later studies. In the Boston Circulatory Arrest Trial however, children with a corrected TGA performed lower than population means on full-scale IQ, *performance IQ*, perceptual organization score, and freedom from distractibility [24]. Children aged 7–11 years with various ConHD showed poorer yet normal intellectual functioning on *full-scale IQ*, *verbal IQ*, *verbal comprehension, and perceptual organization*. Patients aged 12–16 years old showed similar results as healthy peers, with the exception of verbal comprehension [25].

The intellectual functioning of children with hypoplastic left heart syndrome (HLHS) who underwent at least two stages of surgical palliation corresponds to a low average intelligence [26]. Standardized testing in 28 children with HLHS after palliative surgery (mean age at testing 8.6 years) revealed lower performance IQ scores than verbal IQ scores and borderline low-range scores for full-scale IQ in 35.7 % of the survivors, and in 17.8 % IQ scores below 70 were reported [27]. Comparative studies on pre- and postoperative developmental and cognitive functioning in three groups of children (ConHD, children awaiting bone marrow transplantation and healthy children) showed IQ scores within the normal range both before and after surgery. Both the cardiac and bone marrow transplant group, however, had significantly lower IQ scores compared to the healthy group [28].

Thus, undergoing cardiac surgery does not severely impair intellectual function in the majority of the children [29]. IQ scores of the larger part of children with ConHD are within the normal range, although several studies report lower IQ scores in specific groups with more severe cardiac pathology.

5.4.2 School Achievement

Children with transposition of the great arteries (TGA) display overall lower scores on *arithmetic*, *learning*, *and general knowledge tests*. Of 60 children with TGA operated as neonates by means of an arterial switch operation, 23.3 % performed worse than expected on the age of 7 years. In this lower performance group, 18.3 % performed lower than one standard deviation and 5 % performed lower than two SD of the mean score [30]. Children with cyanotic defects in general appear to have lower abilities for arithmetic, reading, and *spelling* [24]. Lower than expected values on reading and mathematics are also found in children with HLHS who underwent staged palliation. Further, even one third of them receive *special education* [this should be Mahle et al. [31]. Although most children with a ConHD seem to perform well at school, about 20 % is considered to perform below average. A study by Shillingford [32] revealed that nearly half of the cohort of children with a ConHD corrected before 2 months of age received some form of remedial services; for 15 % of them this implied full-time special education. This cohort included 46 children with hypoplastic left heart syndrome and other single ventricle deficits following staged reconstruction, towards a Fontan operation; 29 patients with a transposition of the great arteries (TGA) (with or without a ventricle septum defect, VSD) after a arterial switch operation; 12 children with (TOF) after Fallot repair; 12 patients with a VSD with coarctation of the aorta or an interrupted aorta arch; and 11 other ConHD. Mlczoch et al. [33] found the same percentage (15 %) in children with various ConHD (atrial septum defect (ASD), VSD, aortic stenosis, coarctation of the aorta, TGA, TOF, single ventricle pathology) to require special schooling, almost fivefold the percentage found in the overall Austrian background population. No difference in frequency of special schooling was identified within different types of ConHD. The authors stress that a relevant percentage of the children with ConHD will encounter *unemployment* or will need special protected workplaces, which will become a major challenge in the near future.

5.4.3 Attention

Children with TGA, TOF, or VSD were investigated 9–10 years after corrective surgery, and their performances revealed no significant differences on *attention* tasks such as the Stroop Color Word Test and the Trail Making Test, compared to those of healthy children [34]. Another study measuring attention compared children with a secundum atrial septal defect (ASD) that was either surgically closed or by using a transcatheter device [4]. The device group made more errors of commission suggesting *impulsivity*.

The scores on the attentiveness index were also lower. The initial aim of this study however was the comparison of the two treatment approaches and not the investigation of attention problems themselves. At the age of 8 years, children with a corrected TGA by means of an arterial switch operation display a more impulsive response style on a vigilance test. The children assigned to the low-flow cardiopulmonary bypass showed faster reaction times but also made more errors of commission and more anticipatory responses and gave multiple responses compared to those who received total circulatory arrest [24]. Questioning parents and teachers of children with ConHD operated on within the first 2 months of their lives revealed high-risk scores for *inattention* and *hyperactivity* [32]. Using the Attention Network Test, assessing attention networks in the brain, Hövels-Gürich et al. [35] demonstrated poor executive attention skills (*monitoring* and *resolving conflict*) in children with TOF.

Are children with ConHD at higher risk for a diagnosis of *attention-deficit/hyperactivity disorder* (*ADHD*)? In the worldwide pediatric population, the prevalence of ADHD is estimated between 5 % and 7 % [36]. For children with ConHD (the different types were mentioned before) there is a three- to fourfold risk in the prevalence of inattention or hyperactivity [32], and between the ages of 5 and 15 years, the prevalence of ADHD symptomatology was significantly higher in children with both cyanotic and acyanotic ConHD (11.8 %) [37]. In one study on children with HLHS, it was also reported that >50 % of the group demonstrate attention problems [38]. Caution should be indicated for the treatment of patients with ConHD whose circulatory physiology could be compromised by increased heart rates or blood pressure. However, there is the reassuring fact that no clear relationship has been established between stimulant medication used to treat ADHD and the occurrence of sudden unexplained death. The cardiologist and psychiatrist should balance the potential risk for cardiovascular events against the beneficial behavioral effects of the medication [39]. Diagnosing and treating ADHD merits a *multidisciplinary approach*, especially in children with ConHD.

5.4.4 Memory

In 1994, a study including memory tasks in 29 children with TGA or TOF, aged 7–12 years, was reported [40]. All children underwent surgery before the age of 2.5 years. The children with TGA or TOF were compared to 36 children suffering from an innocent cardiac murmur that did not require treatment or a small VSD closing spontaneously. The children performed a number of neuropsychological tests including the *Rey Auditory Verbal Learning Test* and the *Rey-Osterrieth Complex Figure Test*. They did not display any problems on these tasks, and their performances were rated equal to those of the control group. Confirmation of these findings was obtained in other studies in children with TGA [24]. Up to 96.7 % of the children who had an arterial switch operation for TGA (by means of deep hypothermic circulatory arrest or by means of low-flow pulmonary bypass) had a normal performance on learning and memory tasks [41].In children with TOF, we also found lower scores for the patient group in memory for names and narrative

memory [19]. The single ventricle group however displayed significantly scores for design memory at the age of 5 years [42]

5.4.5 Language

Language deficits were observed in children with TGA after neonatal switch operation: 18.3 % displayed reduced scores (<1 SD) on *expressive language* tasks and 21.6 % on *receptive language* tasks (18.3 % <1 SD, 3.3 % <2 SD) [30].

Children with a TGA in combination with VSD operated with use of the total circulatory arrest technique received lower scores on auditory closure and auditory analysis, and a higher percentage of them had an abnormal phonological awareness (59 % versus an expected 34 %). Also *apraxia of speech* occurred more frequently (16 % versus an expected 3 %) [24]. Lower than expected scores on language tests were also found in survivors of staged palliation for HLHS [27], and expressive speech was disturbed in 50 % of patients with (TOF) [43]. Results of our studies also indicate that language remains an area of concern in school-age children with a corrected ConHD. These children have less *phonological awareness* and do not easily gain access to names of color, shapes, and size, and they seem to process instructions less efficiently and less quickly [44]. The deficiency in sound-symbol association and spoken-written word connections might have a negative impact on the acquisition of adequate reading and writing skills.

5.4.6 Psychomotor Functioning

In early research on neuropsychological deficits, reduced *gross motor coordination* in children with cyanotic heart diseases was frequently reported [45], and this finding has been supported by most research to date in various ConHD [19, 34, 40, 44]. Also *fine motor dysfunctions* occur in about 22.1 % of children with TGA after surgical repair [30, 46]. Studying children with both cyanotic and acyanotic ConHDs before and after cardiac surgery revealed poorer locomotor skills both before and after the operation [28]. Gross and fine motor deficits are estimated to appear in 42 % of the children [47].

In our study, we found 25 % of children with various ConHD to perform worse than expected when imitating hand positions and performing subsequent hand movements. Their performances are slower, they display poorer *hand-eye coordination,* and they are less accurate in fine visuomotor tasks [44].

At the age of 8 years, these psychomotor difficulties result in poorer alignment and spacing in handwriting in children with TGA, namely, in those who underwent total circulatory arrest [24]. *Motor slowness* is a major determinant of the psychomotor dysfunctions in school-age children with ConHD awaiting surgery. They take more time to draw connecting lines than healthy controls but also show longer initiation times before drawing the next line. Nevertheless, accuracy is good [48]. Further, the quality of their handwriting did not differ between patients and controls, but more patients

were classified as slow writers. The authors correctly underline the importance of the use of process-oriented tasks in addition to outcome-oriented tasks in studies.

5.4.7 Visual-Perceptual and Visual-Spatial Functioning

Bellinger et al. [49] performed a very elegant study on *visual-spatial skills* in children with TGA with or without VSD after corrective surgery (total circulatory arrest or low-flow bypass). In order to unravel the relative contribution of visual-perceptual abilities, motor control abilities, or metacognitive abilities to visual-spatial (dys) functioning, they included the *Rey-Osterrieth Complex Figure test* (*ROCFT*), as well as the Perceptual Organization Index from the Wechsler Intelligence Scale for Children-3rd version (WISC-III), Grooved Pegboard, and metacognitive abilities. The children (aged 8 years) with TGA and especially those with an associated VSD displayed lower *basal organization scores* on the ROCFT. This lower score was not due to motor control deficits or difficulties with metacognition but reflect visual-perceptual and visual-spatial deficits. In addition, these poor ROCFT scores appeared to be a marker for mathematical deficits and call for early intervention.

5.4.8 Executive Functioning

Bellinger et al. [24] report lower scores on executive functioning in children with a corrected TGA at 8 years of age, especially in *working memory* and *hypothesis generation*. Concerning performances on the Wisconsin Card Sorting Test, the group in total showed great variation, with a higher proportion of them showing lower scores compared to normative data. About 36 % of the children scored lower than 1SD below the mean on number of categories completed and about 25 % on preservative errors (16 % is the expected percentage of children scoring lower than 1 SD below the mean or more in age-expected means). Children get lost in details when asked to perform a task in which they have to structure, pace, or monitor their behavior. While lower-level skills (building blocks, reading individual words) are intact, children display difficulties in the integration and the coordination of these skills to achieve higher-order goals [24]. Calderon et al. [50] also report mild to moderate executive dysfunctions (cognitive and behavioral inhibition) in children with a corrected TGA. Concerning *theory of mind*, the ability to make inferences about people's mental state, children displayed a minimum of 2 years delay, limiting their ability to engage in complex social interactions.

5.5 Summary

Studies in children with various ConHD demonstrate a clear effect of disease severity on cognitive functioning with children with severe ConHD displaying worse cognitive outcome [22]. In early childhood, gross and fine motor problems and

deficits in speech functions and verbal reasoning emerge and to a lesser extent problems with visuospatial skills, number skills, and nonverbal reasoning. In the period of early development, the Bayley Scales of Infant and Toddler Development are essential with the Griffiths developmental scales as an alternative and complementary the Alberta Infant Motor Scale. Research shows that performances on the Mental Development Index of the Bayley Scales at 2 years of age can be predictive for performances at the WPPSI FSIQ at 5 years which validates the importance of early screening.

In the age group of 3–6 years old, motor difficulties and language and visual-motor shortcomings are often reported. A more formal testing can be performed by means of WPPSI, Beery Visual Motor Integration Test, the Peabody Picture Vocabulary Test, and the Movement Assessment Battery for Children, and for a complete testing, we recommend the NEPSY (A Developmental Neuropsychological Assessment). We think it is very important to utilize process-oriented tasks in addition to outcome-oriented tasks in studies, since observation will provide us more information on the cognitive functioning of the child with ConHD and will help us to tailor individual training programs.

The NEPSY is a good clinical instrument that combines both process-oriented and outcome-oriented tasks, and, moreover, the NEPSY is useful in a broad age range (3–12 years). The NEPSY tests the child's neuropsychological development in five functional domains in order to detect subtle deficiencies, within and across these functional domains, which can interfere with learning in preschool and school-age children. In the domain *Attention and Executive Functioning*, children have to perform three tasks. The subtest Tower measures nonverbal planning and problem solving. Auditory Attention and Response Set tests vigilance, sustained auditory attention, and ability to shift and maintain new and complex sets. The subtest Visual Attention reports on speed and accuracy of selectively focusing and maintaining attention on a visual target. The domain *Memory* includes both verbal (Narrative Memory, Memory for Names) and visual memory tasks (Memory For Faces). The subtests Phonological Processing (phonemic awareness), Speeded Naming (access to and production of names of recurring colors, sizes, and shapes), and Comprehension of Instructions (ability to process and respond quickly to verbal instructions of increasing complexity) form the domain *Language. Visuospatial Skills* are assessed by two subtests: Arrows (line orientation and directionality) and Design Copy (ability to copy two-dimensional geometric figures). Fingertip tapping (finger dexterity and motor speed), Imitation of Hand/Finger Positions (ability to imitate hand/finger positions), and Visuomotor Precision (fine motor skills, hand-eye coordination) are the tasks to perform in the domain *Sensorimotor Functioning*. The NEPSY thus combines all cognitive domains and uses very child friendly material that keeps the children motivated.

As the cognitive demand on children increases, more problems supervene during school period and adolescence in communication, handwriting, attention, visual-spatial functions, and executive functioning. These shortcomings may have a negative impact on the acquisition of adequate reading, writing, and arithmetic skills, placing a burden on the child's school career. Intelligence can be tested by means of

Table 5.1 Recommended test material for each age group

Age group (years)	Material	Reference
>3	Bayley Scales of Infant Development	Bayley [51]
	Griffith Development scales	Griffiths [52]
	Alberta Infant Motor Scale	Piper and Darrah [53]
3–6	Wechsler Primary and Preschool Intelligence	Wechsler [59]
	Beery Visual Motor Integration Test	Beery [54]
	Peabody Picture Vocabulary Test	Dunn and Dunn [56]
	Movement Assessment Battery for Children	Henderson and Sudgen [57]
	NEPSY (A Developmental Neuropsychological Assessment)	Korkman et al. [58]
6–18	Wechsler Scale of Intelligence for Children	Wechsler [65]
	Connor's Continuous Performance Test	Connors [55]
	Rey Auditory Verbal Learning Test	Schmidt [63]
	Rey-Osterrieth Complex Figure Test	Rey [62]
	Tower of London	Shallice [64]
	Wisconsin Card Sorting Test	Heaton et al. [60]
	Grooved Pegboard	Reitan and Davidson [61]
	NEPSY (A Developmental Neuropsychological Assessment) (till 12 years)	Korkman et al. [58]
	Assessment by speech pathologist	

the Wechsler Intelligence Scale for Children. In attentional tasks, mainly impulsivity is observed in vigilance tasks, faster working yet more mistakes. The Connor's Continuous Performance Test, Test of Everyday Attention in Children, can be used. Memory seems less involved yet Rey Auditory Verbal Learning Test and the Rey-Osterrieth Complex Figure Test can add important information to the neuropsychological results. Tower of London test and the Wisconsin Card Sorting Test are frequently used in research to test executive functioning. Again in this age group (up to 12 years) I would recommend the NEPSY.

Adequate and recommended test material for each age group is listed in Table 5.1.

Difficult medical circumstances and the acute critical status of the newborn often require immediate medical intervention, reducing the importance of a neurodevelopmental testing in the early stage. Further, the functional testing that can be completed on neonates is extremely limited, and the illness of the child reduces the predictive validity of a preoperative functional assessment. However, it is highly recommended to screen for neurodevelopmental delay in all children with ConHD. Children with mild cardiac symptoms and seemingly normal neurodevelopment should also be screened because several studies proved them not to be free of neuropsychological deficits either [4, 5]. Furthermore, research shows that performances at 2 years of age can be predictive for performances at 5 years which validates the importance of early screening. Because cognitive advancements can be made by intervention provided in the first years of life [1], early identification and subsequently remediation of neuropsychological deficits should be an indispensible part in the treatment of a

child with ConHD. At least should these results prompt cardiologists to thoroughly question the school progress of the child and refer for neuropsychological testing when the risk for neurodevelopmental delay is suspected.

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Part III

Psychological Functioning, Quality of Life and End of Life Care in Adults with ConHD

Psychological Functioning and Life Experiences in Adults with Congenital Heart Disease

6

Edward Callus, Emilia Quadri, Cristiana Passerini, and Anna Tovo

6.1 Introduction

Although it is relatively unknown in the general population, congenital heart disease (ConHD) is the most common inborn defect. It has an approximate prevalence of 8 newborns for every 1,000 births, and this incidence has remained stable for the last 50 years not varying across countries [1].

Not so long ago, these patients used to carry a very poor prognosis especially when it came to the most severe defects; however, nowadays most of them survive reaching adulthood thanks to the advances and triumphs of cardiovascular medicine and surgery in the twentieth century [2]. For this reason, the number of adults with ConHD is growing rapidly; the growth rate has been estimated to be 5 % per year [3].

Due to this, there is an even more clear necessity of understanding of the impact that the pathology and medical condition has on this population and their psychological functioning during childhood and adolescence [4] and also later on during the adult lives [5] so as to develop more personal and humanized processes, systems, and institutions [6].

In this chapter the literature on psychological functioning adults with ConHD will be presented, later focusing on the life experiences and ways of coping of these patients. Finally a clinical case will be proposed, with the objective of clarifying what the role of the psychologist in the context of congenital heart disease could be.

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6.2 Psychological Functioning of Adults with Congenital Heart Disease

When looking at the research available on the psychological functioning of adults with ConHD, there is often a comparison of this group to a healthy comparison or other populations, and the exploration of the correlation of psychological distress with medical and/or other variables.

Although there are common trends, diverging results have also been obtained, and they can be attributed to the vast differences of instruments utilized, but also perhaps to the different health-care systems and health benefits available in the different countries. In this paragraph we especially refer to a recent review conducted in our group [5] in which the labels of psychopathology, anxiety, depression and hostility, neuroticism, and self-esteem were identified.

The studies by Utens et al. and Van Rijen et al. [7, 8] are examples in which there is a comparison between adults ConHD and the healthy peer group. In the study by Utens and colleagues [7], no differences were found between the mean problem scores of the congenital heart disease adults and reference peers. Only some minor, small, and limited differences were found in two specific problem areas: the "Somatic Complaints" and the "Strange" subscales in the study by Utens.

In the second study of Van Rijen et al. [8], it was outlined that young female patients with congenital heart disease aged from 20 to 27 showed higher levels of psychopathology when compared with the reference group; however, when the mean scores of all separate scales of the patient sample and the reference groups were compared, only small differences on the other informant reports and hardly any differences on the self-reports were found.

A study by Cox [9] also confirmed that adults with ConHD had more favorable scores when compared with the control group; however, it is possible that the control group utilized was inappropriate (a group of patients of the department of orthopedics).

In another study of Utens and colleagues [10], a better functioning of the patients with congenital heart disease was found with respect to the reference group, and it was also confirmed that the population had favorable outcomes on daily and leisure time activities. These results were also confirmed in a study by Van Rijen and colleagues [11] in which a better emotional functioning of the congenital heart disease sample is reported when compared to the reference group.

These results indicate that these patients could have a greater sense of coherence [12] with respect to the healthy population, as also outlined in the chapter on quality of life by Apers, Luyckx, and Moons in this book (see Chap. 7). It is indeed possible that having to face an illness these individuals were made to mobilize more resources as they are growing up when compared to the healthy population and search for solutions or ways of coping appropriate to tackle stress [12].

Another possibility that was suggested was that denial and high achievement motivation could inflate the scores on the self-report data [10, 11]. As we shall see further on in this chapter, appearing to be normal is very important for this population, and this further confirms this possibility.

A study which does not confirm this trend is the one by Brandhagen [13], in which it was specified that adults with ConHD fare worse than the healthy population, showing more psychological distress. It is noteworthy that only 168 out of 463 patients completed the questionnaire.

When it comes to anxiety and depression, in a study by Kovacs [14], the objective was to identify the predictors of anxiety and depression in adults with congenital heart disease. It was indicated that 34 % of the patients resulted as having elevated anxiety. Loneliness and fear of negative evaluation, and not variables relating to the medical conditions of the patient, resulted as being predictors of anxiety.

When it comes to depression, 78 % of the sample resulted as having minimal symptoms, 10 % mild symptoms, and 12 % moderate to severe depressive symptomatology. Loneliness, perceived health status, and fear of negative evaluation resulted as being predictors of depression, and the actual disease severity and functional class did not predict mood severity. Similarly, in a study by Rietveld [15], worse scores on psychosocial adjustment and quality of life were linked to having negative thoughts, irrespective of the medical condition of the patients.

Another interesting thing outlined by the study done by Kovacs [14] is that in the subset of their sample who participated in structured clinical interviews, 29 out of 58 patients met diagnostic criteria for at least one lifetime mood or anxiety disorder. Out of these 39 % had never received any mental health treatment.

In a study by Popelova and colleagues [16] on a small group of patients with persistent cyanosis, depression was associated with older age, worse functional state, and unemployment but independent from the severity of cyanosis. In this case 34 % of the sample resulted as being depressed.

Similarly to Kovacs, in the previous studies of Bromberg and Horner [17, 18], psychiatric interviews were utilized to asses adults with ConHD. In the study done by Horner [18] where 29 patients with complex congenital heart disease were evaluated psychiatrically by both interview and questionnaire, the authors confirm that most of the population were functional in day-to-day life; however, 5 were diagnosed as having panic disorder, and 4 patients met full diagnostic criteria for major depression and had received no psychiatric treatment for these conditions.

Also in the study by Bromberg [17], undertreatment seemed to occur as more than one third of the patients who were assumed to be well adjusted experienced a diagnosable psychiatric disorder. Two patients out of 22 met diagnostic criteria for a general anxiety disorder, and 6 out of 22 patients met diagnostic criteria for a depressive episode.

Most of the literature indicates that there is no relationship between diagnosis, physical fitness or presence of residual symptoms, and worse psychological functioning [7–11, 14, 15, 19], with only few studies suggesting a weak link between physical functional class and psychological functioning [16, 20]. For this reason it is particularly important to explore the possible predictors of psychological distress in this population, in order to incorporate them in clinical practice.

6.3 Factors Associated with Psychological Distress

As specified in the previous paragraph, there are strong indications in the literature that the objective physical condition of these patients is not directly linked to their psychological status. In some studies, other possible variables that could influence psychological distress have been explored [5, 14, 15, 19, 20], and these resulted in:

- Loneliness
- Fear of negative evaluation
- Imposed limits
- Low capacity for physical exercise
- Perceived health status

Perceived health status – In our clinical experience, perceived health status is one of the most important factors which influences how the patients feel about themselves, also because it is an indication that there could be significant gaps in illness knowledge in this population [21]. In a recent study in our center, we specifically wanted to examine the effects of perceived health status by including a numerical rating scale assessing the perceived severity of the patient condition [19]. It was verified that the perception of severity of the patient and not the objective medical criteria, including functional class, resulted as being correlated to psychological distress.

Health perception is the result of the integration of the information and feelings related to the health and of one's limits, the health-care system, the family, and the society. It is influenced by childhood experiences, family relationships, working status, and culture. A lot of information is derived from somatic perceptions (symptoms), from the beliefs and knowledge linked to the disease, and from the information arising from the social environment, in particular the ones given by doctors. All of these components entail both evaluations and information both at a conscious level and an unconscious level, together with the resulting emotional responses, and all these processes influence psychological reactions [22].

Low capacity for physical exercise and imposed limitations and restrictions – As specified in the previous paragraphs, although the general trend in the literature is that the more objective medical conditions are generally not related to psychological functioning, in some studies it was pointed out that physical functionality and the more serious conditions have an influence on how the patients feel about themselves. As we shall describe later, it could be that these restrictions and impositions cause problems in daily life activities, causing problems in the social and work spheres of these patients.

Imposed restrictions resulted as being strong predictors of psychological well-being [20, 23, 24], and these are related to physical activity, lifestyle, and to the possibility of pregnancy. These limits may be imposed by the doctor, on the basis of the functional status and the severity of cardiac disease, and they also may be defined by patients themselves on the basis of subjective experiences, or they may be the result of uncertainties and erroneous beliefs. In fact, from our clinical

experience we have seen that it is possible that patients both greatly underestimate the severity of their condition but also that they feel that the severity of their condition is much greater than a more objective medical evaluation of their situation.

Fear of negative evaluation and social isolation – In a review on the biopsychosocial experiences of adults with congenital heart disease in 2005 [25], Kovacs and colleagues outline how congenital heart disease can impact the social sphere of these patients. It is also important to evaluate the qualitative data available [26, 27] in which there is a further indication of the difficulties of these patients, as we shall describe in more in-depth manner later on in this chapter. From our clinical experience and also the data available, there seems to be a struggle to maintain a "normal" life, and this can be hindered from being evaluated negatively by others and also from having the perception that they cannot participate to social and work activities like everybody else. The importance of these factors is highlighted in the study by Kovacs mentioned previously [14] as both fear of negative evaluation and social isolation are linked with depression and anxiety.

6.4 Life Experiences and Coping Strategies in Adults with Congenital Heart Disease

It is very difficult to generalize about the life experiences of this population, since the different conditions are very diverse. Having said this, there seem to be some patterns of experience and perspective in this broad diversity [28]. Having a complex congenital heart condition can be regarded as having a chronic condition because of the long-term nature of the condition, the uncertainty of its course and prognosis, the signs and symptoms of the condition, and also the restriction on the everyday life of these patients [29].

Qualitative studies which take into consideration the life experiences of this population can give valuable insight on how to further improve psychosocial services for these patients. Indications can also be given on how to improve patient compliance and to strengthen the patients' coping strategies by taking into consideration the subjective life experiences of these patients.

In two studies conducted on the adolescent population [30, 31], it was outlined how adolescents with ConHD struggle with physical limitations and face social exclusion. The limitations the patients felt were linked to the severity of the condition, and as many as one fourth of adults with ConHD report their parents as being overprotective during their childhoods and adolescence [13, 30, 32]. This could explain why in general these patients tend to live longer with their parents as overprotection could lead to less autonomy [33, 34]. In both researches the theme of being different and the attempts to cope with this emerged strongly.

This central theme of being different from healthy peers also emerges in the qualitative literature pertaining to the life experiences of these patients in their adulthood [26–28, 35–37], and there are often attempts and the struggle to feel normal and to be perceived as being normal by others which can be followed by feelings of ambivalence and also denial of the condition and efforts to exceed their physical boundaries [35].

In one particular older study [37], the problems encountered by the female patients with ConHD were focused upon mainly the concerns regarding fertility, contraception, and pregnancy. Other concerns which the female patients outlined concerned their surgical scars and body size, and they resulted as being uninformed about aspects regarding their condition and reproductive matters. There is an indication that the perception of body image is quite varied in males and in females and that females experience the consequence of scarring and cyanosis more negatively than men, who seem to have difficulties related to their body image during adolescence [26, 27].

Very often the patients refer to their hospitalization experiences, and in a study [38] it was outlined that the patients, their families, and the nurses had different expectations about the management of the physical needs and about the way the patients and the patients' families behaved during acute phases of the patients' illness. These discrepancies were associated with interpersonal conflict, distrust, anxiety, and dissatisfaction with the health care provided.

As outlined in this manual, the involvement of associations relative to this condition is very important. In the chapters by Geier and Campioni Chaps. 12 and 13, the European experience related to this is outlined. In a very interesting article [28], quotations from patients are reported from the Adult Congenital Heart Association in an attempt to address the patients' perspectives in order to outline the implications for care.

When it comes to feedback regarding their state of health, we find two extremities: on the one hand some were told they were "miracle babies" and also at the other end, lost causes, that they would not survive for long, often erroneously. When told that they were lost causes, this often caused an increase in engagement in highly risky behavior and also difficulties to move into adulthood, because of a perception that there was no future to be put at risk or to work to.

In the conditions of moderate and great complexity, it was reported that there was a risk of language-based misperception in understanding if the condition is completely cured or not. There are also often gaps in knowledge regarding what is required to safeguard and optimize their health [21, 39], and this could be related to the fact that there have not been enough efforts to educate patients regarding their condition, the risks associated with it, and the necessity for lifelong follow-up.

Similarly to our experience, it is common for this population to have difficulties when having to go to the "regular" medical system, where the sanitary personnel is not specialized in ConHD. Not only they need to be vigilant because they might be given medication or treatment which could be inappropriate for them, but they report that insisting for specialized care can result in hostility.

ConHD can be considered as an invisible disability, as quite often it is not evident to others that the person has this condition. The adults often report that friends and family do not really understand their restrictions and that sometimes they also believe that changes in various responsibilities which could be cumbersome are not really required.

Other difficult situations the patients have to face are making difficult decisions regarding their medical care, referral for surgery, and in those occasions when they

are informed that there is nothing left to be done to improve their health [18]. End of life care in this population is a particularly sensitive topic which will be developed in detail by Kovacs and colleagues in this book (Chap. 8).

The patients have also reported that there were occasions in which information is withheld from them and they got to know about their real condition at the onset of new problems. This made it much harder for them to adapt, also because of feelings of betrayal and anger.

Finally, as already specified before, having this condition does not only have negative consequences but there are also some benefits. These include receiving special attention, increased resilience and maturity, and also gaining a clearer sense about the meaning of life. The ongoing awareness of one's mortality can lead to an increased appreciation of life, more clarity of purpose, and better decision making [40].

6.5 Coping Strategies in Adults with Congenital Heart Diseases

From the literature already described, it can be seen that although this population faces many challenges, there is a good social adaptation. In the qualitative study of life experiences already mentioned by Claessens [27], the personality of the subjects is related to the use of particular strategies: in particular, an internal locus of control is related to more active strategies, while adults with an external locus of control seem to be more passive, with a consequent lower adaptation and resignation to feel different, with the consequent risk of social isolation.

In this study, the "process of normalization" is also outlined, understood as dynamic process and management strategy through which the sick person and the family strive to live in a manner similar to that of the healthy peers. This process has its origin from the experience of feeling different from healthy subjects, and it is influenced by the age of the subject and the following factors:

- The environment (ConHD understood as disabilities or realistic vision of the disease)
- The health-care system (medical treatments, approach to the care, and information)
- The personality of the subject

In particular, a realistic view of the disease and the limitations imposed by it, with adequate information, seem to favor the adaptation and coping to the limits imposed by the condition and a better integration of illness management in everyday life.

In a similar study by our group, Callus et al. [26], we also consider the different types of coping strategies utilized to deal with their condition, applying to them the conceptual framework of Moos and Holohan [41]. The patients themselves describe avoiding coping strategies they had utilized previously as not being adaptive,

especially when it comes to the defiance of limits. Other avoidance strategies, such as accepting the inevitable and the utilization of humor, did seem to be adaptive from what the patients described. Denial on the other hand might have been functional at times, such as before an intervention, but it seemed to have consequences anyway, and often it leads the patients not to acknowledge cardiac-related symptoms which could have put their health in danger.

It is interesting to note that the patients who underestimated their conditions the most are the ones who admit ignoring having the condition for many years avoiding medical checkups or defying the therapeutic indications completely. This was also due to the family background they were brought up in and their lack of education and cognitive resources. When dealing with their condition, they describe utilizing mostly cognitive and behavioral avoidance coping strategies.

According to the study of Van Rijen and colleagues [42], the styles of coping of this population resulted as being very similar to their reference groups, with very few differences. The female patients reported lower active problem solving, and the males showed more favorable styles of coping compared to their peers, such as higher seeking of social support, lower passive reaction patterns, and lower expression of negative emotions. Although the adults in this study reported receiving less social support, they also highlighted a lower discrepancy between desired and received social support, indicating feelings of independence in these adults.

In other studies possible defense mechanisms were mentioned [10, 35, 43], in particular the denial of the problems associated with the cardiac malformation and compensation. In particular denial can be manifested in different ways, from the extreme denial of having a condition to the enactment of various strategies to relieve stress. Horner [18] highlights how denial can be adaptive, but that it can also not be functional, worsening the condition of isolation and anxiety.

Strategies for coping in patients with ConHD should be more explored as they are critical in predicting the adaptation to the disease. The education should be customized and designed to replace the responses of passive coping with a more active style of coping, which increases the perception of control over heart condition on the part of the patient and to promote better integration of the disease in the context of life.

In the next paragraph we report a clinical case study of psychological support being provided postoperatively, in an attempt to make a connection between the issues mentioned in the literature and the subjective life experience of one patient.

6.6 The Role of the Psychologist: A Clinical Case Study

Gloria is a 44-year-old woman hospitalized at the IRCCS Policlinico di San Donato Milanese University Hospital for an aortic valve replacement. She is diagnosed with a congenital aortic valvular insufficiency, ascending aorta aneurysm, and pulmonary atresia with an interventricular septum defect.

At the age of 12, she underwent her first cardiac surgery in Birmingham with a positive outcome. Before this surgery, a mild aortic valvular insufficiency was discovered which was followed with regular outpatient visits. At the various controls,

a progressive deterioration of the valve insufficiency and aortic aneurysm dilatation of the ascending aorta associated were discovered, and the patient reported greater fatigue.

At the time of hospitalization, Gloria was referred to the psychological team by the medical staff because she was in a lot of pain due to the fact that she had to keep on the drains for a longer period after the cardiac surgery. The role of the psychologist in psychocardiology is tackled by Prof. Compare and colleagues in this book (Chap. 9), and for these reason here I just give a summary of the role of the psychologist when it comes to this population. Psychologists could be a useful figure in accompanying adults with ConHD patients in the various phases of the illness:

- Coping with becoming aware of the condition or changes in cardiac status [18, 44, 45]
- Accompanying the patient in the difficult pediatric-adult transitions [45, 46]
- Adjustment to cardiac devices and surgical preparation [47–52]
- Maximization of adherence and behavioral modification [53, 54]

It is possible that referrals to psychologists can be initiated by both the patient and the medical staff. It is always important that a psychological (or even a psychiatric) referral is made with the patient's knowledge and to enquire with the patient initially how they feel about receiving this kind of support if it is to be suggested. It is essential to clarify appropriate referral indications with the psychologists working with an adults with ConHD team [45]. Referral to other specialists such as psychiatrists [55] and experts in substance abuse (especially since it has been linked to unsuccessful transitioning from pediatric to adult care [46]) could be necessary in particular cases.

In Gloria's case, she welcomed the possibility to receive psychological support. During the first session, when she is asked how she is feeling, she says that she fears that her reaction to pain and discomfort is not appropriate. When exploring what these feelings are linked to, she explains how everyone she knows, including her husband, encourages her to be strong and to be positive and not to complain and that even if she would like to do so, she is not managing, and for this reason she feels very discouraged.

Firstly, her experience is validated and she is encouraged to talk about how she feels, without passing judgment about this. Gloria welcomes the possibility to be able to express herself, and she also talks about how she felt in some previous hospitalizations.

It is explained to her that it may happen that our loved ones struggle to support our pain, and they try to encourage us. By doing so, they unwillingly do not truly accept and listen to our suffering, and this causes frustration in a person who is in difficulty, who does not feel understood.

Gloria's husband is concerned about her well-being, and he asks the staff about how she is doing, also when it comes to her psychological well-being. It is specified to him that it is quite normal to be experiencing difficulty in this kind of situation, that there are no right or wrong ways to feel and that it is particularly important for cardiac patients who have undergone surgery to feel that there is support without

judgment on how they should be feeling. After this, some indications are given to him on how to be supportive, specifying that it is common to use encouragement in an attempt to help others, but that this can be counterproductive and that it is important in this moment just to listen and to be there for his wife.

In the following sessions with Gloria, the importance of legitimizing one's feelings and giving them relevance is explored – and it is specified that it is an important first step if we also want others to give importance and relevance to what we are feeling and going through. Once she is able to express how she feels regarding her pain and her frustration, Gloria asserts that she feels much better.

The next thing she talks about is her surgical scar. As specified previously, this is an important theme especially for women. She asks feedback about the appearance of her surgical scar and expresses difficulty when it comes to looking at herself. As specified previously, the theme of the surgical scar is important, especially when it comes to females. It is suggested to Gloria to look at herself in the mirror and to express how she feels, welcoming it, while being supported.

She accepts to do this and when she looks at herself, she is very moved. She starts crying and describing on how she has always felt inadequate and that she struggled to accept her illness when she was a little girl. It is suggested to her that the difficulty to look at herself is also linked to these experiences and of seeing herself as being ill; the scar could be a symbol of all of this. Gloria confirms that this is so, and it is agreed that what it means to be ill for her and her feelings of inadequacy are to be explored in the next sessions.

Gloria describes her difficulties at school when she was a child, especially in middle school, the year before she had her first cardiac surgery. She talks about her feelings of inadequacy which were linked to the physical limitations and to the consequent sensations of shame when she compared herself to others in her class. She was particularly ashamed of the fact that she had to be accompanied by her mother in outings because her teachers would not want to take responsibility for her because of her condition. In this case study, it is clearly confirmed that the physical restrictions and the impositions from others have an important impact on the psychological well-being of these patients.

Referring to this in particular, she talks about an episode in which a physical education teacher did not want to include her in the activities because she had this condition and not only – he asks her in front of everyone if she is ill in an indelicate manner. Gloria remembers how she would have liked to become invisible in that moment. She tells everyone to stop looking at her, making the situation worse. Since then she says she will always try to make herself invisible, so as not to be humiliated in front of others.

Remembering about her childhood, Gloria also talks about another physical education teacher, who used to allow her to do physical activity, whatever she managed to do, telling her not to insist if she felt tired. A comparison is made between the two situations, and an agreement is reached about the fact that it is not the condition which made her feel inadequate, but the attitude of some adults who were responsible to take care of her when she was a child which was not appropriate.

After that, it is suggested to Gloria to describe the traumatic event in the present, and Gloria relives the experience and she is very touched emotionally. After that, she is asked to relive the experience as an adult and she said that she would answer back to the teacher. The session is concluded by differentiating these two parts, with the awareness that there is a part of her which feels vulnerable, but also another part which is more adult and which has the capacity to defend itself.

Finally, the possibility that this feeling of inadequacy and shame could have been linked to these memories because they had never been shared with with anyone is explored. In the final two sessions, the themes which emerged are recalled to increase Gloria's awareness on her personal growth. She is very happy to have had the possibility to explore how she feels and her life experiences. She affirms that she probably would not have undertaken psychological treatment, had it not been offered to her in this context. This also confirms the possibility of the undertreatment of psychological distress in this population, as mentioned earlier, which is present in the general population but which could be especially relevant in adults with congenital heart disease, where there is a strong need to feel and seem "normal," often having to face many adversities.

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Quality of Life in Adults with Congenital Heart Disease

7

Silke Apers, Koen Luyckx, and Philip Moons

7.1 Background

7.1.1 The Growing Interest in Quality of Life in Adult Congenital Heart Disease

Advances in medicine, especially in cardiovascular surgery, cardiovascular bypass techniques, and cardiac intensive care, have led to a cascade of improvements for persons with congenital heart disease (ConHD) [1]. *Survival* rates have improved markedly and this has led to an increase in the *prevalence* of adults with ConHD [2, 3]. Consequently, over the past decades, ConHD has been transformed from a disease with lethal consequences into a manageable chronic condition [4]. Many patients with ConHD are not considered to be cured and are more prone to complications resulting from postoperative sequelae or residual defects, which can have a

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considerable impact on their daily lives [5]. Indeed, these patients are now facing long-term psychosocial, neurological, and developmental impairments [6].

As a result of these recent developments in the field of ConHD, health care has been challenged to meet the demands of this complex patient population requiring lifelong care. As such, it has been increasingly recognised that *quality of life* (QOL) has an important role in the evaluation of care and treatment from the patient's perspective. The medical community started showing more *interest* in the concept of QOL since the 1970s and, consequently, QOL was introduced in the database of Medical Subject Headings in 1977. In line with this, the first article to make mention of QOL in ConHD was published by Wennevold and colleagues in 1982 [7]. Ever since, QOL in patients with ConHD has become an important outcome measure in addition to mortality and morbidity. In parallel with the growing consensus that QOL should be measured in patients with ConHD, the number of studies on this topic has increased substantially [8].

One could wonder if QOL deserves this much attention. In other words, is QOL indeed an important factor to focus on in ConHD care and research? With regard to this question, the results of the longitudinal study by Luyckx and colleagues in a large sample of adolescents with ConHD can be enlightening [9]. This study showed that depressive symptoms, loneliness, perceived health, and parental support all uniquely predicted QOL over time, whereas QOL in itself did not predict changes in any of these previous variables [9]. Apparently, QOL was the ultimate *outcome* parameter in this sample of young patients with ConHD. Thus, it is suggested that QOL remains an important outcome measure that deserves continuous attention. This has been confirmed by health-care professionals in the field of ConHD, because they have put QOL high on the research agenda [10].

7.1.2 The Concept of Quality of Life

Despite the growing interest in QOL in adults with congenital heart disease (ACHD), researchers have paid relatively little attention to *conceptual* aspects of QOL [11]. Indeed, QOL in ACHD is often defined by how it is being measured in a specific study (i.e. defining QOL based on the instrument that you use), instead of relying on a thorough conceptualisation for its *measurement* (i.e. choosing an instrument based on how you define QOL). Therefore, a decade ago, Moons and colleagues made a call to perform more conceptually and methodologically rigorous research on QOL in patients with ConHD [11].

Moons et al. evaluated conceptualisations of QOL by looking at their conceptual problems as described by Kinney [11–13]. They found that defining and measuring QOL in terms of satisfaction with life seemed most accurate as this conceptualisation was the only one that dealt with all conceptual problems. Hence, they defined QOL as 'the degree of overall life satisfaction that is positively or negatively influenced by an individual's perception of certain aspects of life that are important to them, including matters both related and unrelated to health' [11]. This definition suggests that QOL is a unidimensional construct (i.e. feeling of overall life

satisfaction) which is influenced by multiple factors (e.g. family or education) [12]. Over the past years, some studies have been published that followed this *definition* and measured QOL in patients with ConHD accordingly [14–19].

In sum, a field of tension between the growing interest in QOL and the methodological problems by which studies are characterised has become apparent. Therefore, the aim of this chapter is to provide a deeper insight into pertinent issues concerning QOL. Firstly, an update on the latest evidence concerning QOL in adults with ConHD is provided. Secondly, pending questions on QOL in ACHD are explored. Thirdly, recommendations for clinical practice are formulated.

7.2 Quality of Life in Adult Congenital Heart Disease: What Do We Already Know?

7.2.1 Recent Evidence on Quality of Life

A complete overview of all studies on QOL in ACHD would lead us too far. Therefore, in this chapter, the latest *evidence* concerning QOL in *ACHD* is provided by focusing on studies published over the past three years (i.e. from 2011 until 2013). In this period, 24 primary studies on QOL in (young) adults with ConHD were published [14–16, 18–38]. Note that studies performed in children or solely in adolescents with ConHD are excluded from this chapter.

The *methods* of the studies published in 2011–2013 are detailed in Table 7.1. The *sample* size of these studies ranged from 12 to 845 patients [26, 38]. However, the majority of studies had samples less than 100 patients (Table 7.1) [14–16, 21, 22, 24–27, 30–34, 36]. Eight different *tools* were used to assess QOL, with The Short Form (36) Health Survey (SF-36) being the most frequently used instrument [22, 23, 28, 29, 32–38], followed by the short version of the World Health Organization Quality of Life Assessment (WHOQOL-BREF) (Table 7.1) [19, 20, 24, 26, 30]. The population studied also varied substantially among the different studies. Most studies, however, investigated patients with a variety of congenital heart lesions (Table 7.1) [18–20, 23–32, 37, 38]. Nearly all studies published in 2011–2013 used a cross-sectional *design* (Table 7.1).

7.2.2 Comparison with Controls

Fourteen of the 24 published studies compared QOL in adults with ConHD to a healthy [14, 21, 33] or community [19, 21–24, 27, 28, 32, 36] *control group* or to other patient populations without ConHD [15, 16]. Four studies found that QOL is worse in patients with ConHD as compared to control subjects [14, 15, 19, 21], whereas three studies concluded that QOL is similar in patients and controls [16, 22, 33]. Further, three studies even found a better QOL in patients with ConHD as compared to a control group [24, 27, 36]. Additionally, three studies examined the scores on different *subscales* of QOL (i.e. SF-36) and found both significantly lower and

Table 7.1 Methods of recent studies on quality of life in adults with congenital heart disease

Study	Sample size	Type of heart defect	Design	Instrument to measure quality of life	Comparison with control group
Chen et al. [20]	$n=289$	Various types of ConHD	Cross-sectional	WHOQOL-BREF	Yes: age-matched general population
Gierat-Haponiuk et al. [21]	$n=30$	Patients who underwent surgery for simple left-to-right shunts	Cross-sectional	EuroQOL-5D	Yes: healthy volunteers
Görler et al. [22]	$n=98$	Patients after surgical repair of transposition of the great arteries (atrial versus arterial switch operation)	Cross-sectional	The Short Form (36) Health Survey	Yes: standard population
Mokhles et al. [23]	$n=463$	Patients with a right ventricular outflow tract reconstruction with an allograft conduit	Cross-sectional	The Short Form (36) Health Survey	Yes: age-adjusted Dutch population norms
Overgaard et al. [14]	$n=62$	Adult survivors with single-ventricle physiology	Cross-sectional	Linear Analog Scale Quality of Life; Satisfaction with Life Scale	Yes: healthy controls
Silva et al. [24]	$n=40$	Various types of ConHD	Cross-sectional	WHOQOL-BREF	Yes: general population
Tay et al. [25]	$n=25$	Patients with cyanotic ConHD and/or the Eisenmenger syndrome	Longitudinal (2 measurements: baseline and after 3 months)	CAMPHOR	No
Tay et al. [26]	$n=12$	Patients with various types of ConHD with Eisenmenger syndrome	Longitudinal (2 measurements: baseline and after 3 months)	CAMPHOR	No
Teixeira et al. [27]	$n=74$	Various types of ConHD	Cross-sectional	WHOQOL-BREF	Yes: Portuguese population as a whole

Vigl et al. [28]	$n=676$	Various types of ConHD	Cross-sectional	The Short Form (36) Health Survey; single-item self-assessment of life satisfaction (ordinal scale from 1 to 7, transformed arithmetically to scale from 0 to 100)	Yes: age- and sex-matched controls of a representative national survey
Cotts et al. [15]	$n=25$	Patients with congenitally corrected transposition of the great arteries	Cross-sectional	Linear Analog Scale Quality of Life; Satisfaction with Life Scale	Yes: adults with mild, haemodynamically insignificant defects
Müller et al. [29]	$n=330$	Various types of ConHD	Cross-sectional	The Short Form (36) Health Survey	No
Opić et al. [16]	$n=26$	Patients with tetralogy of Fallot and an implantable cardioverter defibrillator	Cross-sectional	Linear Analog Scale Quality of Life; Satisfaction with Life Scale	Yes: patients with an implantable cardioverter defibrillator, without tetralogy of Fallot
Schoormans et al. [18]	$n=302$	Various types of ConHD	Cross-sectional	Satisfaction with Life Scale; TNO/AZL Adult Quality of Life-ConHD	No
Bang et al. [30]	$n=85$	Various types of ConHD	Cross-sectional	WHOQOL-BREF	No
Cha et al. [31]	$n=13$	Patients with Eisenmenger syndrome	Longitudinal (2 measurements: baseline and after 6 months)	The Short Form (12) Health Survey	No
Enomoto et al. [32]	$n=72$	Various types of ConHD	Cross-sectional	The Short Form (36) Health Survey	Yes: Japanese normative data

(continued)

Table 7.1 (continued)

Study	Sample size	Type of heart defect	Design	Instrument to measure quality of life	Comparison with control group
Eslami et al. [19]	$n=346$	Various types of ConHD	Cross-sectional	WHOQOL-BREF; Satisfaction with Life Scale	Yes: non-ConHD participants, matched by sex and age
Kahya Eren et al. [33]	$n=69$	Patients with atrial septal defect following percutaneous closure	Cross-sectional	The Short Form (36) Health Survey	Yes: healthy controls matched according to age, sex, educational level, and economic, marital, and employment status
Müller et al. [34]	$n=56$	Patients that had undergone arterial switch operation and atrial redirection (Senning procedure) for transposition of the great arteries	Cross-sectional	The Short Form (36) Health Survey	No
Müller et al. [35]	$n=546$	Various types of ConHD	Cross-sectional	The Short Form (36) Health Survey	No
Ruys et al. [36]	$n=18$	Patients after arterial switch operation	Cross-sectional	The Short Form (36) Health Survey	Yes: age- and gender-specific Dutch population sample
Younge et al. [37]	$n=245$	Patients who had undergone corrective open-heart surgery	Cross-sectional	The Short Form (36) Health Survey	No
Schoormans et al. [38]	$n=845$	Various types of ConHD	Longitudinal (2 measurements: baseline and after 2 years)	The Short Form (36) Health Survey; TAAQOL-ConHD	No

ConHD congenital heart disease

significantly higher scores in patients with ConHD as compared to controls on different subscales of QOL, as well as no differences between both groups for certain subscales [23, 28, 32]. Finally, one study found that women with ConHD had worse QOL than control subjects, while men with ConHD did not differ in terms of QOL from controls [20]. The fact that the results are *inconsistent* could be due to methodological limitations and differences among the studies.

7.2.3 Determinants of Quality of Life

Knowledge of the factors contributing to QOL is of utmost importance in order to improve the effects of therapy and treatment. Sixteen of the 24 studies published on QOL in ACHD examined potential determinants of QOL [14–16, 18–20, 24, 27–30, 33–35, 37, 38]. Table 7.2 provides an overview of the potential *determinants* of QOL that were investigated.

QOL was found to be positively *associated* with having children, higher academic performance or education, higher employment rates or being employed, more daily activities, absence of financial strain, lower employment status, arterial switch surgery, having had cardiac catheterisation, better functional class, better exercise capacity, higher body mass index, better family or social support, being religious, insight into ConHD, lower emotional or psychological distress, extraversion, and sense of coherence (SOC), a concept that will be defined later on in this chapter (Table 7.2) [14, 19, 20, 24, 27–30, 33–35]. Conversely, QOL was found to be negatively associated with cardiac surgery, hospitalisation for ConHD, having an implantable cardioverter defibrillator, physical limitations, somatic symptoms, higher levels of N-terminal pro-brain natriuretic peptide (i.e. a biomarker), type D (distressed) personality, and poor illness perceptions (Table 7.2) [16, 18, 19, 24, 27, 37, 38]. Furthermore, study results are *inconsistent* with regard to the relationship between QOL and age [15, 19, 24, 27, 28, 38], sex [19, 20, 24, 27, 28, 38], medication use [19, 24, 27], disease severity [19, 20, 24, 27, 28, 35], and severity of residual lesions [24, 27] (Table 7.2). Additionally, QOL appeared to be *unrelated* to annual income, ConHD subtype or diagnosis, cyanosis, time of diagnosis, ConHD illness course, personal resources, and family environment (Table 7.2) [19, 24, 27, 30].

Apparently, studies published over the past three years identified more *determinants* of better or worse QOL than factors that are unrelated to QOL. However, it should be noted that several of these potential determinants were investigated in only one study. When having a closer look at the potential determinants of QOL that were examined in more than one study, patients' age and sex were the most frequently examined determinants, followed by academic performance or education, disease severity, cardiac surgery, functional class, medication, and social support. Factors that were consistently associated with a better QOL were higher academic performance or educational level, better functional class, the absence of prior cardiac surgery, and better social or family support (Table 7.2). For the other factors, the findings were inconsistent.

Table 7.2 Overview of potential determinants of quality of life in adults with congenital heart disease

	Chen et al. [20]	Overgaard et al. [14]	Silva et al. [24]	Teixeira et al. [27]	Vigl et al. [28]	Cotts et al. [15]	Müller et al. [29]	Opic et al. (2012)	Schoormans et al. [47]	Bang et al. [30]	Eslami et al. [19]	Kahya Eren et al. [33]	Müller et al. [34]	Müller et al. [35]	Younge et al. [37]	Schoormans et al. [38]
Demographic characteristics																
Older age			↗	=	↘	=					↘					↗/↘
Female sex	↘		=	=	↘						↗					↘
Having children											↗					
Higher academic performance or education				↗	↗						↗	↗				
Higher employment rate or being employed					↗						↗					
More daily activities							↗									
Absence of financial strain											↗					
Lower employment status											↗					
Annual income											=					
Clinical characteristics																
Cardiac surgery			↘	↘							↘					
Surgical strategy (arterial switch compared to Senning)													↗			
Cardiac catheterisation											↗					
ConHD hospitalisation											↘					
Medication			=	↘							=					

Better functional class	↗	↗														↗
More severe heart defects	↘		=	=	↘						=			↘/↗		
More severe residua			↘	=												
ConHD subtype or diagnosis				=						=						
Cyanosis			=													
Implantable cardioverter defibrillator								↘								
Physical limitations				↘												
Better exercise capacity							↗									
Time of diagnosis											=					
ConHD illness course										=						
Higher BMI											↗					
Having somatic symptoms											↘					
Higher levels of N-terminal pro-brain natriuretic peptide															↘	
Psychosocial characteristics																
Personal resources			=													
Family environment			=													
Better family or social support	↗		↗	↗							↗					
Type D (distressed) personality									↘							
Being religious										↗						

(continued)

Table 7.2 (continued)

	Chen et al. [20]	Overgaard et al. [14]	Silva et al. [24]	Teixeira et al. [27]	Vigl et al. [28]	Cotts et al. [15]	Müller et al. [29]	Opic et al. (2012)	Schoormans et al. [47]	Bang et al. [30]	Eslami et al. [19]	Kahya Eren et al. [33]	Müller et al. [34]	Müller et al. [35]	Younge et al. [37]	Schoormans et al. [38]
Insight into ConHD										↗						
Lower emotional or psychological distress	↗										↗					
Poor illness perceptions																↘
Extraversion	↗															
Sense of coherence														↗		

↗ potential determinant of better quality of life, ↘ potential determinant of worse quality of life, = unrelated to quality of life
ConHD congenital heart disease, *BMI* body mass index

The use of a *cross-sectional* design in most of these recently published studies permits investigation of associations between potential determinants and QOL, but does not make it possible to draw conclusions in terms of directionality of effects. For that purpose, *longitudinal* studies are required. To date, only one longitudinal study has been published that examined the direction of effects between QOL and potential determinants [9]. This study was conducted in adolescents with ConHD (age 14–19 years) and investigated the direction of effects between QOL and both contextual and individual resources (i.e. parental support, peer support, and SOC), accounting for the role of perceived health status and controlling for sex, family structure, and disease complexity. This study indicated that perceived health, parental support, and SOC predict changes in QOL over time [9].

In summary, evidence is mixed with regard to the level of QOL in adults with ConHD compared to controls. Furthermore, QOL in adults with ConHD seems to depend on multiple factors and is to a limited extent only determined by their heart defect. Higher academic performance or education, better functional class, absence of prior cardiac surgery, and better social and family support are consistently associated with a better QOL. However, the comparability of the study findings is hampered due to differences in the research methods.

7.3 Quality of Life in Adult Congenital Heart Disease: What Do We Still Need to Know?

Irrespective of the plethora of QOL studies in ACHD, there are still several issues that we need to know. In this respect, it is important to first summarise the *gaps* in the current knowledge base and, subsequently, to provide avenues for future research.

7.3.1 Gaps in the Current Knowledge Base

Firstly, there is no consensus on the definition of QOL and its measurement in the field of ACHD. This lack of a clear *conceptual* background has led researchers to use QOL to describe a range of different variables. As a result, the instruments used to measure QOL vary greatly [39]. Remarkably, the *SF-36* was the most frequently used instrument to measure QOL in studies published during the past three years, although this instrument is generally assumed to measure patients' physical and mental health status [40]. Apparently, researchers do not make a clear distinction between health-related QOL and QOL. The term *health-related QOL* is often used although, in reality, perceived health status is being measured [40]. In addition, previous research demonstrated that QOL and health status are two distinct constructs that cannot be used interchangeably [9, 41, 42]. Therefore, researchers ought to rely on sound conceptual grounds to choose the most appropriate instrument for measuring QOL.

Secondly, most studies use a *cross-sectional* design which makes it impossible to draw valid conclusions about temporal sequences and the direction of effects [39, 43]. However, this is crucial information for the design of effective *interventions* aimed at improving QOL. Furthermore, the use of a cross-sectional design makes it impossible to look at long-term changes in QOL. As such, the potential effects of life-changing clinical events on QOL (e.g. surgery) or the effects of an intervention aiming to improve outcomes cannot be evaluated in the long run.

Thirdly, adults with ConHD constitute a heterogeneous group of patients. The impact of ConHD varies significantly as a result of the great diversity in anatomical complexity and disease course. Therefore, some studies focused on a specific *subpopulation* of patients (e.g. patients with the same diagnosis or patients who underwent a specific type of surgery). However, in order to obtain a complete overview of QOL in ACHD, more information is needed on patients from the whole spectrum of heart defects, accounting for their type of ConHD but also for disease progression, symptoms, and so forth. It would be interesting to extensively examine the potential influence of the ConHD subtypes and to compare QOL among patients with different types of heart defects.

Fourthly, QOL is likely to differ between *countries*, each with their own health-care system and sociocultural environment [44]. However, the bulk of recent studies on QOL in ACHD are conducted in Western countries. Moreover, there is a lack of studies comparing QOL between countries. Hence, it remains unknown how patients with ConHD from around the world are doing in terms of QOL and how their QOL relates to country-specific characteristics such as the organisation of health care.

These gaps in the current knowledge base show that it is difficult to interpret recent results. Furthermore, due to methodological differences and shortcomings, inconsistent study results are the connecting thread in QOL research in ACHD. Hence, to guide clinical decision-making and health policy, it is crucial to continue investing in high-quality *research* and to bear in mind that there is still a lot to learn about QOL in ACHD.

7.3.2 Avenues for Future Research

It is currently not possible to draw firm conclusions about the level of QOL and determinants of QOL in this patient population. It has been recognised that true excellence in care cannot be achieved without excellence in research [45]. Therefore, more methodologically and conceptually sound research is urgently needed.

Table 7.3 gives an overview of important avenues for future research. In general, three types of research are suggested to expand our knowledge base of QOL. Firstly, *longitudinal* research is required to examine how QOL changes over time and to assess the direction of effects between QOL and possible determinants and concomitants in adults with ConHD. Detailed information on change and stability of QOL can guide the identification of vulnerable groups of patients having, for example, a consistently low QOL over time. Secondly, *multicentre* studies in an *international* context using one well-defined method to measure QOL are needed. Such a research strategy will ensure the generalisability of the findings and will enable researchers and health-care professionals to make valid comparisons. In addition,

Table 7.3 Recommendations for future research on quality of life in congenital heart disease

Longitudinal research is needed to:	Examine changes in quality of life over time
	Assess the direction of effects between quality of life and possible determinants and concomitants
Multicenter, international studies should be initiated using one common, well-defined method to measure quality of life in order to:	Be able to make valid comparisons
	Gain insight in whether the reported discordant results are due to methodological differences or whether they represent real differences, for example, between countries or regions
	Examine which factors impact on differences in quality of life
	Ensure the generalisability of the findings
Intervention studies could be conducted aimed at improving modifiable determinants of quality of life such as:	Psychosocial and personality factors

this will help us to understand whether the inconsistent research findings are due to methodological differences or whether they represent real differences, for example, between countries and regions. Such studies should also examine which factors impact on differences in QOL worldwide. Thirdly, a critical step in future research is to understand how QOL can be optimised in patients with ConHD through the implementation of *interventions* that target modifiable determinants. Such interventions should be tailored to the needs of specific populations and increased attention should be paid to vulnerable patients. Therefore, health-care professionals should be aware of the factors that influence QOL in patients with ConHD, such as social support or certain personality characteristics [18, 24, 27, 46].

In addition to moving on to specific types of research, there are specific topics that should be elaborated on as well. For example, the use of biomarkers and the genetic basis for QOL are upcoming issues in the field of ConHD. With regard to the use of *biomarkers*, recent study results showed that N-terminal pro-brain natriuretic peptide (i.e. a marker for heart failure) was related to patients' physical QOL [37]. Furthermore, individuals' *genetic* predisposition was recently put forward as a potential explanation for the variation between patients in terms of QOL in individuals with Marfan syndrome, which cannot be explained by other patient characteristics [47]. More specifically, Schoormans et al. found a genetic basis for mental QOL in cytokine-related genes and their activity [47]. These promising results should be backed up by additional studies examining the predictive value of biomarkers in ConHD and validating findings on cytokine genetic pathways in larger samples.

7.4 Quality of Life in Adult Congenital Heart Disease: What Do We Learn from This?

Of course, health-care professionals ought to be aware of the (fixed) factors that influence patients' QOL, such as sex. However, the key for improving QOL is to identify modifiable *determinants*. In doing so, health-care professionals should be

aware of the fact that not only medical variables play a role in patients' QOL. Indeed, some *psychosocial* factors have a major impact on QOL in addition to, often unchangeable, demographic (e.g. age) or clinical (e.g. cardiac surgery) characteristics. After identifying such modifiable determinants, caregivers should aim to improve patients' QOL through tailored intervention efforts. Therefore, we formulate several recommendations for clinical practice to guide health-care professionals in supporting QOL in ACHD.

7.4.1 Modifiable Determinants of Quality of Life

Two examples of psychosocial factors that are particularly actionable are patients' SOC and their illness perceptions [38, 48]. From a clinical perspective, such characteristics can be of great importance to support QOL.

Indeed, *SOC* is a variable of particular interest as a determinant of QOL [9]. SOC is the core construct of the *salutogenesis* theory, which has been developed in line with the emerging paradigm of positive psychology [49, 50]. Salutogenesis concentrates on the origins of health and well-being, which is in contrast to the paradigm of *pathogenesis* looking at the origins of disease and ill-being [49]. SOC represents an individual's generalised world view and expresses the extent to which the individual perceives: (1) stimuli as structured and predictable (i.e. comprehensibility), (2) that resources are available to meet the demands posed by these stimuli (i.e. manageability), and (3) that these demands are challenges worthy of investment (i.e. meaningfulness). In fact, the salutogenic model seeks to describe the processes through which people manage to stay healthy despite being exposed to ubiquitous stressors. It is theorised that SOC exerts its positive influence on health through adaptive health behaviours and coping behaviour [49].

Extensive reviews of the literature confirmed the relation between SOC, psychosocial health, and QOL in diverse patient populations and individuals from the general population [51, 52]. Furthermore, it has been hypothesised that patients with ConHD may develop a strong SOC as their feelings of comprehensibility, manageability, and meaningfulness (i.e. the three components of SOC) may be bolstered by disease-related experiences from a young age, such as openly discussing their concerns with health-care professionals or receiving structured education [53].

So far, six studies on SOC in adolescents with ConHD have been published [48, 54–58]. Two of these studies compared SOC of patients with healthy peers and concluded that SOC in adolescents with ConHD was significantly higher as compared to healthy peers [48, 54]. Furthermore, cross-sectional studies found that *SOC* was associated with anxiety and depression, perceived health, and QOL [48, 57, 58]. Longitudinal studies concluded that SOC predicts QOL and perceived health [55, 56]. Hence, the current evidence seems to confirm the hypothesis that patients with ConHD develop a strong SOC and that SOC is a pathway to improve their QOL. To date, only two studies were published on SOC in adults with ConHD [14, 35]. These studies concluded that SOC scores in patients with single-ventricle

physiology did not differ from those of controls, whereas SOC was enhanced in patients with various types of ConHD compared to reference values [14, 35]. Furthermore, SOC was also associated with QOL [35]. More research on SOC in adults with ConHD is required.

In addition, *illness perceptions* may also be a promising *determinant* of QOL in ACHD and, thus, a possible target for interventions to increase QOL [38]. Illness perceptions are defined as the cognitive representations and beliefs that patients have about their illness [59]. These illness perceptions consist of six components, namely, (1) personal consequences or the effects on physical and psychosocial functioning, (2) personal and (3) treatment control or the potential of modifying the illness and its effects through personal or treatment interference, (4) illness coherence or patients' understanding of their illness, (5) timeline cycle or the perceived changeability of the illness and its effects, and (6) emotional representations or the experience of negative emotions as a result of the illness [38]. However, more research is needed in the field of ConHD as only one study investigated the predictive value of illness perceptions on QOL in ACHD [38].

7.4.2 Overview of Possible Intervention Programmes to Improve Quality of Life

The next step in improving QOL is to design *interventions* targeting modifiable determinants. In theory, improving patients' QOL could be established, for example, through enhancing patients' *SOC* and changing their *illness perceptions*. Indeed, based on the current evidence in adolescents with ConHD, SOC seems to be a key factor in the promotion of QOL. There are, however, no studies examining interventions strengthening SOC and, consequently, improving QOL in patients with ConHD. Such interventions should focus on empowering patients and enhancing their feelings of comprehensibility, manageability, and meaningfulness by providing information, making patients aware of available resources, and supporting patients to adopt an active role in their daily care [53]. In daily clinical practice, health-care professionals could anchor medical encounters around these three *actionable* components of SOC.

In order to increase QOL, health-care professionals could also aim to improve the different components of patients' illness perceptions. Examples of interventions that proved to be effective in other cardiovascular populations are informing and educating patients to increase their knowledge of the disease (i.e. illness coherence) and providing cognitive behavioural therapy to improve patients' proactive thinking about their illness [38, 60].

To date, there are no intervention studies (i.e. randomised controlled trials) on the improvement of QOL in ACHD, despite the growing interest in QOL. Researchers and health-care professionals should join forces to move on from merely observing patients' QOL to improving their QOL. Certain psychosocial determinants appear to be suitable targets for intervention as they are modifiable and highly actionable.

Conclusion

ACHD is a challenging and evolving field in terms of QOL. Therefore, this chapter aimed to provide an update on the recent evidence with regard to QOL in ACHD. A large variability among studies assessing QOL was observed. More specifically, results are mixed regarding the level of QOL in patients with ConHD in comparison to control subjects and there is no consensus on which *determinants* are important towards QOL. The inconsistent research findings are largely due to methodological differences and limitations of the respective studies. Hence, the current body of knowledge does not provide conclusive evidence on QOL in adults with ConHD and, furthermore, does not allow for designing prevention or *intervention* programmes that boost QOL. Recent study results have suggested that the detrimental effects of ConHD on the daily lives of patients can be compensated by *psychosocial* factors, such as patients' illness perceptions. However, we still need to know more about QOL and gain in-depth understanding of its modifiable determinants. To achieve this goal, more *longitudinal* studies and studies performed in an *international* context based on a solid conceptual framework are needed. Indeed, the biggest challenge related to QOL in the field of ACHD is to find conceptual and methodological consensus and, subsequently, to move on from observational studies to interventional research. To meet this challenge, researchers and health-care professionals should team up in order to bridge the gap between living longer and living well in patients with ConHD.

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End-of-Life Care and Treatment Preferences Among Adults with Congenital Heart Disease

8

Meaghan K. Ferguson and Adrienne H. Kovacs

8.1 Introduction

Due to significant developments in cardiac surgery, pediatric cardiology, and medical technologies, individuals born with congenital heart disease (ConHD) are now living longer than ever before [1]. Approximately 85 % of infants born with ConHD now survive to reach adulthood, which is in stark contrast to the much lower survival rates of only a few decades ago [2]. Experts estimated that there were approximately 800,000 adults living with ConHD in the United States in 2000 [3]. Despite notable medical advances resulting in increased life expectancy, the long-term prognosis for this population remains of concern [4]. Adult patients with defects of moderate or great complexity remains at risk of premature death from progressive heart failure or arrhythmias [5–10] and/or a prolonged period of progressive disease before death [4, 7, 10]. For many patients, cardiac surgeries are considered palliative measures rather than curative ones because they do not modify the basic anatomic or physiologic disturbance [11, 12]. For these reasons, as a group, patients would likely benefit from advance care planning (ACP) and palliative or end-of-life (EOL) care similar to that which is recommended for other chronically ill patients, such as those with cancer or heart failure associated with acquired heart disease [13, 14].

Guidelines for ACP and EOL care are more readily available for patients with heart failure associated with acquired heart disease than for patients with heart failure due to ConHD [15, 16]. Published American guidelines for the management of adults with ConHD recommend that all patients complete advance directives, but do not otherwise mention ACP or palliative care [12]. It is thus advantageous to draw upon guidelines for the care of adults with heart failure due

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to acquired heart disease. As such, comprehensive care should include the assessment and management of both physical and psychosocial symptoms, attention directed to health-related quality of life, and discussion regarding EOL preferences and caregiver support [5, 16].

Emerging research provides valuable insight into the needs and preferences of adults with ConHD as they prepare for the end of their lives. This chapter will begin with a summary of the current mortality expectations of individuals born with ConHD. Next, there will be an overview of what is currently known about ACP and EOL care for adults with ConHD, including a discussion of both barriers and facilitators. This chapter will conclude with clinical recommendations, with a focus on the potential roles of psychologists and other mental health professionals.

8.2 Changing Mortality Patterns

There has been a noticeable decline in the mortality of patients with ConHD as evidenced by a number of cohort studies. For example, Billet et al. examined the national mortality rate of patients in England and observed a 26 % decrease in age-standardized mortality rates for ConHD between 1994 and 2003 (collapsed across all age categories) [17]. Boneva et al. observed a similar reduction in mortality (39 %) between 1979 and 1997 based on data available from the multiple-cause mortality files compiled by the National Center for Health Statistics from the Centers for Disease Control and Prevention [18]. In a Canadian evaluation of adults with ConHD in Quebec from 1987 to 2005, mortality decreased by 31 % across the last 5 years of the study (2002–2005) compared to the earlier study years (1987–1990) [2]. Improvements were most prominently influenced by the reduction of childhood deaths among infants born with more severe forms of ConHD [2]. A large retrospective study conducted by Pillutla et al. utilized information from the Centers for Disease Control Multiple Cause-of-Death Registry to determine trends in mortality from 1979 to 2005 among adults with ConHD in the United States [1]. Results from the study were positive such that there had been a decline in mortality since 1979 for almost every type of lesion, including tetralogy of Fallot, transposition of the great arteries, atrial septal defect, ventricular septal defect, patent ductus arteriosus, and congenital aortic stenosis. The diagnosis and management of ConHD has changed significantly over the past 40 years, such that the number of adults living with severe ConHD defects now rivals the number of children living with the same [19]. Marelli et al. found that, between 1985 and 2000, the prevalence of severe ConHD in adults increased by 85 %, whereas it increased by only 22 % among pediatric populations [19]. This further illustrates the increasing presence of survivors with complex ConHD who will require long-term care as they continue to age.

In the past, research investigating the mortality associated with ConHD focused on pediatric populations and there was less known about the causes of death in adult patients. In line with the changing life expectancies of this population, recent research has more thoroughly investigated the changing causes and

rates of mortality of adults with ConHD. A cohort study evaluating the 25-year mortality rate of adults with ConHD in Oregon in the United States observed that mortality from surgical procedures had declined, although late mortality associated with cardiac complications remained high [20]. It is now known that adults with ConHD die from a range of both cardiovascular and non-cardiovascular causes. Among non-cardiovascular deaths, malignancy and pneumonia are the predominant causes [4]. Since 2000, for adults with non-cyanotic forms of ConHD, myocardial infarction has become the leading cause of death, suggesting that coronary heart disease is developing within this aging population [1]. Arrhythmia and heart failure, however, remain the primary contributing causes of death for cyanotic lesions [1]. Whereas death due to heart failure is typically a prolonged and progressive course, sudden cardiac death also affects approximately one-quarter of patients [4, 10].

Verheugt and colleagues analyzed the Dutch CONCOR national registry for adult ConHD, which is linked to the national mortality registry, and reviewed 6,933 patients [4]. Of this sample, 197 (3 %) died during the total follow-up of 24,860 patient years. The majority of the deceased patients were male (58 %) and the median age at death was 49 years (range: 20–91 years). On average, patients died much younger from sudden death (39 years) than heart failure (51 years). Defects with the highest mortality were univentricular complexes, double outlet right ventricle, and tricuspid atresia. Further, endocarditis, supraventricular arrhythmia, ventricular arrhythmias, conduction disturbances, myocardial infarction, and pulmonary hypertension were each associated with an increased risk of cardiovascular or non-cardiovascular mortality (after adjustment for age, gender, and defect complexity). This study highlighted that, despite improvements in survival, patients with ConHD have excess mortality compared to the general population and that the vast majority of patients die from cardiovascular causes.

There is thus a growing cohort of adult survivors with complex ConHD who will require specialized care as they continue to age. Adults with complex forms of ConHD are considered to have a chronic medical condition associated with a shortened life expectancy, with many experiencing a prolonged state of illness prior to death [5]. Although patients now live longer, cardiac complications such as arrhythmias and heart failure often appear later in life. Surgical and transplant options may be limited to a select subset of patients [21]. Therefore, as individuals with ConHD progress into adulthood, disease management may be considered palliative, such that it entails a shift from a primary focus on life-prolonging measures to increased attention targeting enhancement of quality of life. As such, adults with ConHD may benefit from the same palliative care principles that are regularly applied to other chronically ill groups of patients, including those with acquired heart disease [13, 16]. Comprehensive palliative care involves ACP and EOL discussions between patients, families, and health-care providers. Among adults with heart failure associated with heart disease and other groups of chronically ill adults, ACP and EOL discussions have been associated with positive outcomes [22, 23]. There is now a small but growing body of literature that is investigating current practices of ACP and EOL care among adults with ConHD.

8.3 Advance Care Planning for Adults with Congenital Heart Disease

Advance care directives are written documents that allow individuals to name a substitute decision maker and identify treatment and health-care preferences in advance of a time (e.g., when they are seriously ill or dying) when they may be incapable of communicating for themselves. Such documents typically benefit patients, family members, and health-care providers. Previous research in congestive heart failure has shown that patients prefer to have information about their medical condition and prognosis communicated to them in a timely fashion prior to the onset of debilitating stages of illness [11, 23, 24]. Early knowledge and information about the possibility of death from progressing illness may be helpful in later communication and care planning when the patient's health is deteriorating and it is more difficult to plan [25]. The phrase "hope for the best, and prepare for the worst" has been recommended for communicating the importance of ACP [25]. Given the chronic nature of ConHD, the known common sequelae (e.g., arrhythmias and heart failure), and the increased risk for premature death, ConHD represents a lifelong illness that requires both active care and future planning. The 2008 American College of Cardiology/American Heart Association guidelines for the management of adults with ConHD recommend that patients complete ACP directives "at a time during which they are not morbidly ill or hospitalized, so that they can express their wishes in a less stressful setting" [12].

The completion of advance care directives, however, does not appear to reflect standard practice among adults with ConHD [26, 27]. Tobler and colleagues evaluated the prevalence of the completion of advance directives by patients as well as patient preference regarding life expectancy information [26]. Two hundred outpatients with ConHD (mean age = 35 ± 15 years; 52 % male), of whom the majority (81 %) had moderate to severe disease complexity, completed surveys assessing their knowledge and preferences for advance care planning. Of the total sample, only 5 % had completed any advance directives and over half (56 %) indicated that they had never heard of this term. Despite these findings, a substantial majority of the sample (87 %) reported that it would be important to have an advance directive in place should they be dying and unable to express their wishes. Further, in the event that they were to become too unwell to make independent health-care decisions, most patients indicated a preference for family members (86 %) rather than physicians (50 %) to make their health-care decisions. Although only 35 % of patients reported that they would like to know when they would die (if that information was available), double this proportion (70 %) were interested in receiving information about the general life expectancy of adults with their same form of ConHD. In summary, as a group, adults with ConHD appear interested in completing advance directives, engaging family members in health-care decision-making, and obtaining information about longer-term health-care expectations. These goals can be achieved if discussions about advance care planning become a routine practice, as has recently been recommended [12, 28].

8.4 End-of-Life Care of Adults with Congenital Heart Disease

The patient perspective regarding EOL matters was explored in a survey of 123 adults with ConHD regarding their level of concern on a scale ranging from 1 ("not at all concerned") to 5 ("extremely concerned") on various medical, psychosocial, and lifestyle topics [29]. The mean score on the item labeled "death and dying" was 2.8 ± 1.3, with 57 % endorsing moderate to extreme concern (defined as a score of between 3 and 5 on the 5-point scale). When contemplating their own death and dying, the following items were identified as being "very important" by more than half of the respondents in Tobler et al.'s survey of adults with ConHD: (1) honest answers from doctors, (2) understanding treatment choices, (3) visits from family and friends, (4) having said everything they want to say, (5) being free from pain, (6) being physically comfortable, (7) not being a burden to loved ones, (8) knowing how to say good-bye, (9) being at peace spiritually, and (10) fulfilling personal goals/pleasures [26].

Tobler et al. found that 78 % of patients thought that a member of their medical team should initiate EOL discussions and the majority also agreed that the cardiologist was the most appropriate provider to do so [11]. The preference for EOL discussions did not differ by any demographic variables or disease complexity. In contrast to this reported preference to have EOL conversations, only 1 % of patients recalled having EOL discussions with their caregivers. Tobler et al. also surveyed 48 health professionals (primarily cardiologists) providing care to adults with ConHD [11]. Fifty percent of this provider sample reported that they commonly initiated discussions of life expectancy, advance care planning, and resuscitation preferences. Both patients and providers were asked the same question: "When do you think is the best time to bring up end-of-life discussions?" Whereas the majority of patients were in favor of earlier discussions (i.e., "before getting sick"), most providers thought that these discussions should wait until patients are first diagnosed or symptomatic with a life-threatening complication. These findings draw attention to the infrequency at which EOL discussions with adults with ConHD actually occur (or are recalled by patients) as well as the discrepancy between patient and provider perceptions regarding the optimal timing of such discussions.

The two aforementioned studies included surveys of patient preferences and recalled EOL discussions. There is a notable contrast between 1 % of patients recalling EOL discussions with providers and 50 % of providers saying that they commonly initiate such conversations. One possible explanation is that patients do not identify conversations regarding life expectancy, advance care planning, or resuscitation as EOL discussions [11]. Indeed, studies with other medical populations revealed that patients often have difficulty understanding EOL discussions when they do occur [30]. While this may also be the case with adults with ConHD, the results of another study suggest that EOL discussions with adults with ConHD are not regularly documented. Specifically, researchers retrospectively examined the experience of 48 patients with ConHD who died in hospital due to cardiac causes (mean age at death 37 ± 14 years) [27]. Careful medical chart abstraction was

performed in order to document the circumstances of death, EOL discussions, and the provision of EOL care. Most patients in this study cohort had defects of moderate to severe complexity and 42 % had previously undergone assessment for heart transplantation. In other words, death was not completely unanticipated. Heart failure was the documented cause of death for 50 % of patients. Prior to death, 44 % of patients were on mechanical ventilation and 15 % were undergoing dialysis; 52 % of patients died under attempted resuscitation. Only 10 % of the patients, however, had documented EOL discussions either before or during their final admissions. Documented EOL discussions with either patients or their substitute decision makers were associated with a significantly lower incidence of attempted resuscitation (12 % vs. 100 %). Further, not one patient who had been referred to palliative care died under attempted resuscitation.

8.5 Barriers and Facilitators to Effective Advance Care Planning

In summary, both providers and patients recognize the importance of advance care planning for adults with ConHD [11, 12], yet such planning does not seem to occur with regular frequency [26, 27]. It is thus important to identify possible barriers and facilitators for advance care planning and the initiation of EOL conversations. The optimal time at which to initiate EOL discussions, for instance, may be unclear and even challenging [21]. The chronic nature of ConHD often means that there is not always a defined trigger for deterioration of health [21]. Therefore, knowing when the most appropriate time to engage in EOL conversations may be difficult [21].

From the aforementioned cohort of 200 adults with ConHD and 48 health-care providers, Greutmann et al. investigated the most commonly endorsed barriers and facilitators to EOL discussions [5]. With regard to barriers, 85 % of providers expressed concern about their ability to reliably estimate the life expectancy of adults with ConHD. Other concerns endorsed by over half of the providers included the belief that patients with life expectancies greater than 10 years may be unprepared for EOL discussions (79 %), the possibility of negative patient reactions to EOL discussions (65 %), worry that the patients' wishes may change over time (56 %), and the notion that the initiation of EOL discussions may take away patients' hope (52 %). Patient concerns that might be considered barriers to EOL conversations include a preference for concentrating on staying alive rather than talking about death (66 %) and uncertainty regarding what type of care they would want to receive should they become seriously ill (55 %).

With regard to facilitators to EOL communication, Greutmann et al. reported that providers' willingness and engagement in EOL discussions were positively impacted by having a good patient-provider relationship (96 %), having known the patient for a long time (83 %), and confidence in the patient's prognosis (83 %) [5]. The most common patient-reported facilitators included trust in the physician (85 %), confidence in the physician's ability to treat ConHD (78 %), and the belief that the physician will be available should the patient become very sick (69 %) [5].

Tobler et al. also noted that the two most important factors for adults with ConHD when contemplating issues of death and dying were genuine, honest responses from physicians and an understanding of treatment options [26].

In summary, factors that impact the occurrence of EOL discussions, both positively and negatively, pertain largely to the quality of the patient-provider relationship as well as the availability of information regarding medical treatment options and prognostic expectations.

8.6 Clinical Recommendations

The initiation of discussions regarding ACP and EOL issues should not be avoided despite the challenges of prognostication. For adults with ConHD of moderate or great complexity, given the chronic nature of the disease, the risks of prolonged illness and sudden death, and the shortened life expectancy, ACP and EOL discussions should become a standard component of ConHD care. There indeed exist challenges regarding prognostication, but such challenges can actually be integrated into discussions. For example, providers can inform a patient about what is currently known about the likely medical sequelae and life expectancy associated with their particular form of ConHD, while at the same time acknowledging (a) the limitations when applying information to an individual and (b) the fact that published data might reflect patients managed in a former era when newer interventional or medication strategies were not available. Patients themselves seem more interested in general information regarding life expectancy rather than predicting their own age of death [26]. Discussions pertaining to longer-term outcomes and care strategies should begin from an early age, even prior to transfer to adult care. This is already practiced with patients with other pediatric-onset illnesses, such as cystic fibrosis, and research suggests that the quality of palliative care is enhanced when discussions begin earlier in the discourse of the disease [31]. As outlined earlier, adults with ConHD also favor earlier initiation of EOL discussions at a time when they are relatively healthy [24]. It has been recommended that such discussions be "normalized" with patients and considered an appropriate part of ongoing medical care [28].

Professional education regarding effective patient-provider communication should be offered in order to support effective ACP and EOL discussions. Both patients and providers acknowledge the importance of establishing and maintaining rapport when discussing matters related to EOL care [5]. Instructing and/or reinforcing skills for empathic and open communication may improve providers' skills for relaying mortality information or advance care options. It is also important that providers ensure that patients recognize EOL discussions when they do occur [11]. The term "life-shortening medical condition" has been recommended [28]. Research suggests that physicians and trainees who participate in emotional skills training perform better with interpersonal communication than those who have not been part of such training [32, 33]. There is even evidence to suggest that ConHD providers are keen for such skills training. Specifically, in their survey of 48 ConHD providers, Greutmann et al. found that 79 % were interested in receiving more

information and resources regarding advance directives and 89 % were interested in ACP and EOL communication strategies [5]. The "Ask-Tell-Ask" framework may provide guidance for EOL discussions [28, 34]. Within this approach, providers may (1) ask what a patient understands about the situation (e.g., prognosis, EOL care options), (2) tell information that is requested or should be communicated, and then (3) ask whether the information was understood and whether the patient has any further questions.

Ongoing patient education should remain a priority. Patients may lack information about the potential course of their illness, future possibilities for active treatment, and/or EOL care options. Therefore, in order to make advance care decisions, patients must first be thoroughly educated regarding the anticipated illness trajectory, the benefits of ACP, as well as the EOL care options that will be made available to them. A proactive approach is recommended [21]. The creation of standardized information, available in both written format as well as on the Internet, would allow patients to access important information and prepare questions to ask their healthcare providers at future medical appointments.

Increased collaboration with palliative care experts should be sought. There is room for improvement in collaboration between physicians, patients, and palliative care professionals [35]. A partnership between ConHD providers and palliative care colleagues may be optimal in order to comanage patient care [28].

8.7 The Role of Psychologists and Other Mental Health Professionals

The experience and expertise of psychologists can provide a valuable asset within a multidisciplinary ConHD care team [36]. Although the term psychologist is used throughout this section, the roles described may also apply to other mental health professionals including psychiatrists, counselors, and clinical social workers.

Supporting the needs of health professionals taking care of adults with ConHD. Open and honest lines of communication are critical to facilitate effective ACP and EOL discussions [26]. Cardiologists generally receive minimal formal training in communication skills and/or the psychological aspects of living with chronic medical conditions. Psychologists, on the other hand, generally have thorough, specialized training in assessment, case conceptualization, and individual, family, and group psychotherapy. Incorporating psychologists into the discussion may be beneficial given their practice with using emotional language, discussing sensitive matters such as death, and identifying patient emotions and reactions [28, 36]. Psychologists may also provide professional education opportunities for physicians who may wish to improve communication of issues related to death and dying to both patients and grieving family members. Additional professional training may include the psychosocial and ethical aspects of death and dying as well as the consideration of cognitive-developmental and cross-cultural issues.

Supporting the needs of adults with ConHD. Health psychologists in particular have specialized training to understand the interaction between physical and

psychological functioning and to foster coping and resilience for individuals living with (and dying from) chronic medical conditions. Psychologists may help adults with ConHD through the provision of emotional support, an open discussion of anticipatory grief, and a review of strategies to communicate with loved ones and health professionals [36]. Published guidelines for the care of adults with ConHD recommended that consultation with psychology or psychiatry is indicated to "facilitate optimal functioning" to improve adjustment to worsening illness or hospitalizations [37]. Physicians are not alone in their discomfort with conversations about death and dying. Many patients may also have difficulty discussing EOL issues, particularly if their health is already deteriorating. It can be discouraging and frightening for patients to experience declines in their functional health status or to discuss the possibility of failing health and eventual death. For patients struggling to cope with the impact and implications of EOL discussions, psychologists may provide emotional support and strategies to manage health- and mortality-related psychological distress. Patients having difficulty when faced with different ACP/EOL options (e.g., cardiac device deactivation) may also benefit from consultation with a psychologist who can support effective decision-making [36].

Supporting the needs of families of adults with ConHD. As a group, adults with ConHD wish their family members to be an integral part of ACP and EOL discussions [26]. A multidisciplinary team that includes psychologists might serve to facilitate these discussions and enhance the quality and understanding of the conversation as perceived by patients as well as family members. Parents often play a very important role in the lives of young adults with ConHD who are facing the end of their lives. Throughout a patient's infancy and childhood, parents are responsible for medical decision-making and often continue to play a major role during adulthood. Parents' involvement in ACP and EOL planning should be commensurate with the stated wishes of the patients. Following a patient's death, psychologists may also work with bereaved family members.

Contributing to interdisciplinary research. As psychologists with doctoral degrees receive extensive training in research design and statistical analysis, psychologists may offer a unique perspective within interdisciplinary care teams [36]. They are well poised to contribute to the growing body of research exploring the preferences and experiences of adults with ConHD with respect to ACP and EOL as well as the development of educational and supportive interventions targeting both patients and providers.

8.8 Summary and Conclusions

The majority of patients born with ConHD will now survive to reach adulthood, though adults with defects of moderate or great complexity remain at particular risk of cardiac complications and premature mortality. Emerging research suggests that, as a group, adults with ConHD wish to discuss EOL issues with their medical team and prepare advance planning documents. Although there is a noted discrepancy between patient preferences and usual clinical practice, there is reason for optimism

given providers' reported interest in learning about advance directives as well as strategies to improve EOL communication. Providers are encouraged to consider EOL discussions as an important component of comprehensive ConHD care and also to be mindful of the importance of a strong patient-provider relationship when patients first begin thinking about, and later actually face, the end of their lives. Psychologists and other mental health professionals are well suited to support the needs of patients, their family members, and health-care providers with the shared goal of enhancing both quality of life and the quality of the death experience.

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Part IV

Psychological and Peer to Peer Interventions and the Importance of Non-profit Associations

Psychocardiology and the Role of the Psychologist in Acquired and Congenital Heart Disease

9

Angelo Compare, Cristina Zarbo, and Annalisa Bonaiti

9.1 Psychocardiology

Psychocardiology is a field that refers to the role of clinical psychology in cardiac setting, and it is related to the biopsychosocial factors which emerge in the onset, the course, and the rehabilitation of a heart disease.

The traditional separation between medicine and psychology has gradually reduced during the years, thanks to the acknowledgment of the disease as a complex interaction of different factors (individual characteristics, biological processes, and social experiences) and consequently to the increasing number of psychologists who work with physicians and other health-care providers. The several changes in psychological paradigms (behavioral, cognitive behavioral, psychodynamic, systemic, phenomenological, and so on) have influenced the applications of clinical psychology to medicine [1]. One of the turning points has certainly been the introduction of the biopsychosocial model presented by George Engel [2], which has contributed to change radically the approach to the disease and to the patient. Health psychology has been applied in the *acquired* cardiac setting since some decades.

Many psychological factors influence the course of cardiac disease, such as depression and anxiety. For this reason, psychological and cardiac issues cannot be seen separately. It is necessary, instead, to take on a multidisciplinary approach because clinical psychology may contribute to prevent, understand, and treat the cardiac disease. Clinical psychologists have to tailor their interventions to the specific needs of cardiac patients, which necessitate a lot of support because of changes in their quality of life due to frequent hospitalization and difficulties at work and in

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their social sphere [3]. However, psychological and cardiological components do not have to be considered as a single unit; the different competencies of each professional need to be emphasized in order to work at the same level to improve the prevention and the treatment of the cardiac disease [1].

Psychocardiology has a very long tradition. In both the clinical tradition and in common sense, there is an attempt to try to find an association between psychological, social, and emotional factors with cardiac disease [1]. Evidence of this association can be found since 1628, thanks to William Harvey who claimed that "mental agitation" influenced the heart's functioning. However, the systematic study of the relationship between cardiac disease and psychological factors began in the 1950s with the research of Friedman and Rosenman about the "type A behavior pattern," which was found to be a factor generally present in cardiac patients. After that, other studies continued to focus on investigating this relationship. In the 1960s, it was found that some addictive behaviors, such as smoking and alcoholism, and health-related lifestyle were associated with cardiac disease, and for this reason, cognitive behavioral programs aimed to modify these risk factors were organized [4–7]. Today, cardiac disease may be considered as a complex pathology that reflects the realities of living in modern society, such as stress, lack of time, competitiveness, and excessive ambition, which are all part of the dynamic of success in contemporary cultures [8].

More recent studies found an association between cardiac disease and psychopathology, especially concerning anxiety, depression, excessive workload, stress, and social isolation. Furthermore, a relationship between psychosocial factors and the occurrence and course of cardiac disease was found [9].

In cardiac settings, psychologists have to deal with behavioral, emotional, and relational problems which affect patients and often also their family. According to Molinari and colleagues [1], there are different areas that should be considered by a psychologist who works with cardiac patients:

1. Symptomatology
2. Physical functionality
3. Psychological functionality
4. Disease history
5. Disease perception/processing
6. Resource, coping, self-efficacy
7. Familiar and social support
8. Motivation toward therapy and attitude with respect to adherence
9. Expectations

Furthermore, the specific needs of cardiac patients and the relationship established with the medical team and the psychologist need to be considered. It has been suggested in the pertinent literature that the whole history of the life of a person and the family context in which he or she lived needs to be known to take care of the patient [10].

The role of the psychologist in cardiac settings changes depending on the selected clinical approach. Molinari and colleagues [1] investigated four main psychological approaches which could be applied in the treatment of a cardiac disease.

1. According to the idea of "patient-focused therapy" [11], the patient has the necessary resources to reach the individual well-being and psychology has to assume a nondirective stance to increase therapy compliance.
2. The studies of *positive psychology* emphasize the role of positive experience of life and individual resources and abilities. In cardiac settings, positive psychology focuses on the promotion of quality of life for patients and their families.
3. *Cognitive behavioral therapy* tries to change the dysfunctional behaviors in the "hic et nunc" of the therapy activating patients' skills and valid strategies to overcome problems. The cognitive behavioral therapy is particularly useful for the treatment of cardiac patients because it is a brief intervention and therapist and patients work actively to identify specific cognitive pattern that may generate problems. Furthermore, it helps to reduce risk factors, such as smoking and unbalanced diet, and also to increase self-efficacy, self-esteem, and the internal locus of control [12].
4. *Systemic family therapy* is particularly indicated to deal with the crisis of the patient and his family after the occurrence of a cardiac event. Due to the disease, the family system and its balance structure break down and the roles have to be renegotiated. Systemic family therapy helps patients to manage with the emotional distress and conflicts which may emerge and affect the whole family's quality of life and patient's medical and psychological conditions.

9.2 Psychological Issues in Acquired and Congenital Heart Disease

9.2.1 Acquired Heart Disease

The World Health Organization [13] has reported that coronary heart disease causes approximately 7.2 million deaths every year. Literature suggests that several psychosocial factors such as distress, anxiety, type A and type D trait personality, depression, worries, loneliness, and low social support may influence acquired cardiac disease morbidity and prognosis and affect cardiac patients' quality of life [14–20]. In particular, depression and anxiety are the primary risk factors for adverse outcomes in several cardiac populations. Both direct (biological) and indirect (behavioral) mediating processes explain the negative effect of psychological issues on cardiac disease. The biological mechanisms include inflammatory and immune processes, alterations in activating HPA, variability in heart rate, increased activity of the sympathoadrenal and pituitary–adrenal axes, reduction in circulating endothelial progenitor cells, increase of cortisol and catecholamine levels, alteration of

activities of the autonomic nervous system, and oxidation processes. Moreover, unhealthy lifestyles are often linked to depression and anxiety; no medical adherence; increased consumption of tobacco, alcohol, and illicit substances; reduced physical activity; and overeating that may contribute to a worse prognosis of cardiac disease. Moreover, it has been shown in the literature that there is a nonlinear and simultaneous pathway between traditional risk factors for cardiac disease and personality traits [21]. A further psychosocial risk factor for cardiac disease morbidity and mortality is low, or the absence of, social support. Evidence has in fact shown that a lack of social support may lead to negative psychological states like anxiety or depression which, in turn, may influence health through direct effects on physiological processes or through adverse health behaviors [22]. Social support may contribute to health status through protecting the subject from the potentially pathogenic influence of stressful events (the buffering model) or providing positive experiences and stability in life situation (the main-effect model) [23]. It has been in fact demonstrated that low quality of couple relationship emphasized the negative influence of depression on cardiovascular rehabilitation outcomes [24].

In a single case study, it emerges that cardiac disease is related to a profile of cognitive processing of emotions characterized by elevated levels of unprocessed emotions, low appraisal, and high suppression emotional regulation strategy [25]. Also takotsubo syndrome has been proposed to be linked to cognitive emotional processing [26].

9.2.2 Congenital Heart Disease (ConHD)

Annually, approximately 0.5–1 % of infants are born with a congenital heart disease (ConHD), that is, a heterogeneous group of infants with abnormalities affecting the heart and associated vessels. ConHD is now one of the most common congenital malformations in the world [27]. In recent years the expectations of life after surgery for ConHD are increasingly improved, and this has led researchers to pay more attention to the impact that it has on the psychological and cognitive functioning of the child/adolescent and their families.

9.2.2.1 ConHD Patients' Psychological Issues

ConHD patients face a variety of biopsychosocial issues that may lead to premature mortality, neurocognitive deficits, body image concerns, physical and cognitive limitations, lower educational and occupational attainment, delayed social maturation, and limitations in interpersonal relationships. Moreover, the persistence of severe or disabling physical health problems into adulthood may increase the possibility of later psychiatric difficulties [28].

The current literature suggests that the presence of ConHD may impact intellectual and other neurocognitive abilities negatively. Children and adolescents with ConHD report in fact high rates of learning disabilities and reduced both cognitive functioning and school performance [29–34]. Moreover, they are at greater risk of poorer intellectual functioning and developmental difficulties such as visual/perceptual, motor, and speech/language skills than healthy children.

Children with chronic disease present moreover the highest risk for emotional and behavioral problems [35, 36]. Areas of functioning that may be particularly affected include neurocognitive functioning, body image, social and peer relationships, and mild delays in developmental functioning [37]. In some cases, it has also been observed the presence of eating and sleep disorders [38]. Patients with ConHD often report low self-esteem [39], which would be considered a personal resource that facilitates a positive perception of stressful life situations and reduces psychological stress.

However, scientific literature underlines that the only presence of ConHD does not adversely affect behavioral problems among children and adolescents. Exposure to potential risk factors during the course of a patient's life may increase the development of specifically internalizing problems in older children and adolescents with ConHD [40]. Literature suggests that ConHD patients exhibit more internalizing than externalizing problems [36, 41].

One of the most frequent internalizing problems is heart-focused anxiety, which can be defined as a fear of heart-related symptoms and sensations precipitated by perceived negative consequences associated with cardiac-related sensations. Heart-focused anxiety is significantly correlated with increased symptoms of anxiety and depression and lower health-related quality of life among patients undergoing cardiac surgery [42]. Health-related quality of life (HRQoL) includes the patient's subjective, emotional evaluation, reaction to problems, and limitations and is influenced by a variety of different biological and psychosocial determinants. ConHD patients report significantly poor quality of life, especially on motor functioning, cognitive functioning, autonomy, social functioning, and positive emotional functioning [43]. Two main groups of factors may contribute to developmental outcomes in ConHD patients. Firstly, there are medical and procedural factors, such as the duration of surgery, hospital stay, and characteristics of special support techniques during surgery. Secondly, there are familial and environmental conditions, like parental concern, resources, and support. Successful adaptation to disease depends on adequate self-regulation between biological and familial components.

It has been suggested that adults with ConHD may show difficulties in several areas of functioning, including neurocognitive functioning, body image, social and peer relationships, and mild delays in developmental functioning [37]. In adults with ConHD, loneliness, fear of negative evaluation, imposed limits, low capacity for physical exercise, and perceived health status are the main predictors of psychological distress [44].

Moreover, there are indications that the condition of diversity is the core of the emotional experiences connected to ConHD in adult age. Feeling different and being perceived as being different are interlinked and coping strategies adopted resulted as being influenced by this negative perception [45].

9.2.2.2 ConHD Families' Psychological Issues

The experience of childbirth and becoming a parent can become distressing when newborns have serious chronic illnesses such as a ConHD. It has been shown in the scientific literature that parents of ConHD children experience psychosocial morbidity to a higher degree than parents of children with other pediatric conditions and parents of healthy children [46]. The excessive care burden, coupled by other

detrimental factors related with parenting a child with severe chronic illness, is in fact likely to affect familial well-being and quality of life.

It has been shown in literature that parents of children with ConHD experience more fear, anxiety, depression, hopelessness, and distress compared to parents of children with other diseases and parents of healthy children. The parental crisis may be reflected on marital couple, and partners may tend to isolate themselves, creating a fracture inside and increasing marital tension [47]. The reaction to the disease, however, is determined both by what it evokes and triggers in both parents and the quality of the preexisting relationship.

Negative mental health consequences are most evident among mothers [48–50], possibly because they are the primary caregivers in most families [29]. Mothers experience in fact more stress, guilt, and sadness if compared with fathers [51] and develop more symptoms of anxiety and depression [52]. However, maternal well-being depends on the severity of the child's ConHD and the time of measurement. Less severe and shorter-term stressors, like having a child with mild or moderate ConHD, do not lead to lower levels of overall well-being [53]. It has been shown that it is essentially the burden of caregiving and the presence of the disease within their social environment (the so-called family effect) that affect both the physical and mental health of the caregiver [54]. There is moreover an association between increased maternal anxiety/protection and poor emotional adjustment in children with ConHD. Parental overprotection, which can be defined as an excessive contact, infantilization, and prevention of independent behavior, may be a risk factor for the pathogenesis of anxiety disorders [55] and might contribute to increased heart-focused anxiety in ConHD patients through selective learning and reinforcement of "cardio protective" behaviors. Overprotective parents may hold in fact negative outcome expectancies about their child's ability to cope with stressful events and therefore discourage child's participation in potentially stressful activities, which may in turn encourage avoidance behaviors.

It is recognized that the quality of infant–parent relationship can moderate mental disease risk and that good parental mental health and availability to respond to the child will improve psychological and physical outcomes in the child.

It is suggested in the scientific literature that a positive perception of the disease allows to affect psychosocial outcome positively and, on the other hand, negative perceptions (such as distressful, demanding, etc.) may affect health outcome negatively [56–59]. Parental well-being is an important indicator of positive mental health and might act as a protective factor against mental disorders for both parents and children.

9.3 Psychological Interventions for Acquired and Congenital Heart Disease

9.3.1 The Role of the Psychologist

Guidelines and recommendations to deal with a cardiac disease identify mental health as an important factor as patients are considered to be at increased risk of psychosocial, emotional, and behavioral difficulties, and these issues may be taken into account in intervention and care programs for patients and their families.

According to the American Heart Association, cardiac rehabilitation refers to multidisciplinary interventions designed to optimize the activities of physicals, psychologists, and medical team, with the aim of reducing morbidity and mortality [60]. The psychologist is a professional qualifying element of interdisciplinarity that allows to give a comprehensive response to the patient. The psychologist plays a very important role in helping the medical team inside and outside the hospital to offer support to patients and families on several issues such as therapeutic adherence, adjustment of lifestyles, and reworking the trauma related to the disease. Unfortunately, rehabilitation programs for ConHD patients today are almost nonexistent, and psychologists play a key role only in pre- and postsurgical time. For this reason, there is the need to support ConHD patients and families and plan specific rehabilitation programs that include the psychological aspects.

The psychologist also plays a key role in the personalization of the therapy. It has been shown that medical optimization, understanding of relational and professional pressures which collides with the patient daily, and the anticipation of emotional difficulties and the consequent impact on the physical level facilitate the patients' adherence and decrease the rates of hospitalization due to relapses. Psychologists can educate medical staff with regard to the importance of effective communication and patient participation in their medical care [61], which leads to a differential diagnosis that takes into account the specificity of each individual patient.

The psychological intervention will aim to help patients and their families to recognize and express their emotions regarding the disease, identify and implement strategies for the control of risk factors and modification of lifestyle, implement self-management of rehabilitation treatments on the basis of individual characteristics, and, finally, regain a satisfactory quality of life.

The disease is a critical event that can destabilize the family system and can induce changes all inside of it. A good cardiac rehabilitation should then include the patient's family and investigate their cognitive aspects, that is, the level of awareness that family members have about the disease and the attribution of meaning given to it; emotional aspects, that is, the emotional impact produced by the disease on the members of the family and the degree of support that they offer; and relational aspects, that is, the style of interaction between components of the family, changes in relationships and roles that can result from illness, and eventual loss of autonomy by the patient [62].

9.3.2 Psychological Interventions

9.3.2.1 Psychological Interventions for Acquired and Congenital Heart Disease Patients

Evidence suggests that cognitive behavioral therapy is one of the most effective interventions for cardiac patients with anxiety and depression [63]. In cognitive behavioral therapy, patients are taught to restructure anxiety-provoking thoughts leading to panic attacks, are taught relaxation techniques to counteract stress and anxiety, and are given exposure therapy to desensitize themselves to stressful stimuli. This therapy conveys the message to the patient that it is possible to learn

self-management techniques and methods to self-control. Cognitive behavioral therapy may provide a suitable treatment option for grown-up congenital heart survivors to reduce risk factors and unhealthy behaviors. This approach could be used to challenge unhelpful core beliefs (e.g., about being abnormal or different) that may underline problems with low self-esteem and unhelpful coping strategies such as avoidance or the use of alcohol or drugs.

A Rogerian client-centered counseling could facilitate adjustment and supports clients for difficult decisions. This approach could also facilitate the processing of surgical interventions and other difficulties such as grief and loss. Resilience and an increased empathy may be considered the major benefits of early adversity.

Interpersonal psychotherapy (IPT) could offer a further treatment option. A focus on the psychosocial challenges that patients are facing may prove more beneficial effects than focusing on intrapsychic properties which may risk pathologizing a normal reaction to challenging life events. IPT is an empirically validated, time-limited intervention (12–16 sessions) that focuses on symptom reduction and improving the quality of the client's social and interpersonal functioning. IPT is based on the premise that an interpersonal crisis triggers psychological distress. The interpersonal triad models the development of this distress. In addition to an interpersonal stressor, the triad comprises the client's biopsychosocial vulnerabilities (including genetic vulnerability, temperament, attachment style, and personality) and social circumstances. IPT could be beneficial for adult survivors of cardiac illness by validating the psychosocial challenges that they face, linking these life events to affect and endeavoring to find a social network that better meets their emotional needs.

Moreover, it has been shown that identifying and reinforcing social support networks and its quality may be helpful to improve adherence behaviors and quality of life, in particular of patients with depressive symptoms. Including diet therapy, exercise training and psychological counseling in cardiovascular programs seem to be relevant for the effectiveness of the treatment. It has been shown in the literature that a multidisciplinary CR program is effective on functional exercise capacity, BMI, and general well-being outcomes [64].

Finally, latest evidence suggests that complex intervention can be facilitated by the use of telemedicine that allows the remote control of considerable amounts of clinical data (e.g., ICAROS project) [65, 66]. A consistent preference by ConHD patients for psychological treatment to be provided over the Internet vs. in person or over the telephone has been shown in a study [67]. It has been demonstrated that well-being therapy (WBT) based on personalized mobile technology allows therapist to support patients' well-being in real life and in a continuous way [68]. Also the home blood pressure telemonitoring (HBPT) has been found to be effective in improving blood pressure control and related medical and economic outcomes in hypertensive patients with metabolic syndrome [69].

However, although many studies on the psychological treatment of cardiac patients have been published, a randomized controlled trial is needed to assess the effects of psychological interventions for depression in congenital heart disease [70].

9.3.2.2 Psychological Interventions for ConHD Families

The modern paradigm of care for children with serious congenital heart disease requires a multidisciplinary approach to care that involves their families. A better and accurate diagnostic information from the cardiologist during the pregnancy, associated with anticipation of the management of the child, should be promoted in the areas of obstetric care, neonatal care, and intensive care. It would be appropriate to provide also guidelines to give information on the management of the situation in the case of specific medical interventions. It has been found that a lack of information can lead parents to feel distrust, greater distress, and a sense of lack of control. Data on pediatric care indicate that parents want to be involved in their children's treatment and rehabilitation.

ConHD families need to call for "Family Enrichment" treatments, which are psychological interventions of preventive–promotional nature that focus on the family resources, have the aim of enhancing specific skills of the couple and parents, and improve its functioning and prevent problematic developments [71].

The Congenital Heart Disease Intervention Programme (CHIP), based on a "transactional" stress and coping model of understanding child outcomes in chronic illness, places the appraisal and coping style of the family, and especially the mother, in a central position as potentiating, or protecting against, the inherent stresses posed by the illness context [72]. The CHIP is a comprehensive and brief program of early psychosocial interventions for mothers of infants with congenital heart disease which has three main aims:

1. To help parents to construct meaning and process the grief associated with the loss of the anticipated "normal" child, through narrative therapy processes
2. To promote effective mother–infant transactions related to feeding, caretaking, and social and sensory stimulation
3. To act on maternal coping skills against worry, anxiety, and stress and to teach mothers active, generalizable, problem-solving strategies to address current worries related to having a child with congenital heart disease

The CHIP interventions were deemed to be as beneficial to infants with, as well as without, associated developmental syndromes. They were shown to have beneficial effects on infant neurodevelopment, feeding transactions, maternal appraisal, worry, and anxiety [73].

Conclusion

In summary, psychological factors have a great impact on acquired and congenital cardiac disease etiology and influence its course (see Table **9.1**). For this reason, they have to be considered in the cardiac setting, especially when they are related to acquired and congenital heart disease. Cardiac patients may be affected by many biopsychosocial issues related to the emotional functioning, and also their families can experience more psychological problems compared with the family of children with other disease or with family of healthy children. Therefore, psychologists play a key role in helping the medical team to support patients and their

Table 9.1 Topic aspects about psychocardiology and the role of psychologist in acquired and congenital heart disease

1. *Psychocardiology*
Psychocardiology is a psychological field which refers to the role of clinical psychology in the cardiac settings, and it is related to the biopsychosocial factors which emerge in the onset, the course, and the rehabilitation of a heart disease
In cardiac setting, psychologists have to deal with behavioral, emotional, and relational problems which affect patients and often also their family. They should consider several areas such as symptomatology, physical and psychological functionality, disease perception/processing, resource, coping, self-efficacy, familiar and social support, motivation and medical adherence, and expectations
The role of the psychologist in cardiac settings changes depending on the selected clinical approach. The most common clinical approaches are patient-focused therapy, positive psychology, cognitive behavioral therapy, and systemic family therapy
2. *Psychological issues in acquired and congenital heart disease*
Several psychosocial factors such as distress, anxiety, type A and type D trait personality, depression, loneliness, worries, and low social support may influence acquired cardiac disease morbidity and prognosis
Both direct (biological) and indirect (behavioral) mediating processes may explain the effects of psychological issues on cardiac disease
Congenital heart disease patients face a variety of biopsychosocial issues that may lead to premature mortality; neurocognitive deficits; body image concerns; physical, interpersonal relationship, and cognitive limitations; highest risk for emotional and behavioral problems; eating and sleep disorders; low self-esteem; heart-focused anxiety; and lower health-related quality of life (HRQoL)
Parents of children with congenital heart disease, especially mothers, experience fear, anxiety, depression, hopelessness, distress, overprotection, and marital tension. Parents' well-being might act as a protective factor against mental disorders for both parents and children
3. *Psychological interventions for acquired and congenital heart disease*
Evidence suggests that cognitive behavioral therapy is one of the most effective interventions for cardiac patients, especially with anxiety and depression. Also Rogerian client-centered counseling and interpersonal psychotherapy (IPT) may be beneficial for both acquired and congenital cardiac disease patients
ICT technologies may allow psychologist to assess and support cardiac patients in real life and in a continuous way
The modern paradigm of care for children with congenital heart disease requires a multidisciplinary approach to care that involves families of ConHD child. Family Enrichment treatments and Congenital Heart Disease Intervention Programme (CHIP) are the most common interventions for ConHD families
Psychologists play a key role in helping the medical team to support patients and their families in the onset, the course, and the rehabilitation of a heart disease

families in the onset, the course, and the rehabilitation of a heart disease. Furthermore, psychologists can provide health-care practitioners information about the effectiveness of medical interventions by exploring how the patients feel and also help clinicians determine the best ways to manage heart disease [3].

Further research should continue to investigate the psychocardiology field to find the more appropriate solutions and interventions in order to facilitate a good recovery and improve the general health status of patients.

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10 Communication and Relational Aspects in Healthcare Professionals Caring for Pediatric and Adolescent Patients with Congenital Heart Disease and Their Families

Maria Simonetta Spada, Lorenzo Galletti, Valentina Strappa, and Angelo Compare

The pentagram of interpersonal motivational systems can produce endless 'relational tunes,' through varied sequences and combinations of its 'notes,' just like musical notes, allowing an infinite range of compositions.
Liotti

10.1 The Plurality of Meanings, Social Representations, and Symbolic Values Related to the Heart

The heart is an organ like no other. It has long been used as a symbol related to spirituality, emotions, and morality, inherent in the human being. In the past it was believed to be the place of the human mind, and, even today, the word heart continues to be used poetically to refer to the soul, and stylized heart shapes – not exactly corresponding to its real form – are widely used as symbols of love. "Love and gentle heart are one thing" ("Amore e cor gentil sono una cosa"), Dante wrote in *Vita Nuova*, 1293–1295.

Having a cardiac illness means not only to be affected in a vital organ but also in the vital center of energy itself. When we think about CPR, we usually figure it as an attempt to restore energy and to keep or bring the person back into the world of

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the living. In our clinical practice, a child with a pacemaker to stimulate the contraction of his/her heart said to his/her mother: "You know mum, I am like those robots that work with batteries: if I run out of energy I stop."

Even in mithology, where the body with its functions prevails over mental contents (such as emotions, torment, psychic pain), the heart maintains a privileged role "[…] the heart is stirred inside the breast […]" [1]. Chest pain can thus assume a communicative dimension, that is, it narrates its inner world and becomes a means of symbolic communication of emotions.

Jean-Luc Nancy [2] writes: "from the moment I was told that I needed a transplant (of the heart), all signs seemed to falter, all references capsized (…) Just the physical sensation of an already existing void in the chest, a sort of apnea in which nothing, absolutely nothing, not even today, would be able to separate myself from the symbolic and the imaginary, nor to separate the continuous from the interrupted (…) it was one feeling: that I had fallen into the sea while still remaining on the bridge."

Bringing your hands to your chest indicates the precordial constriction, and to the neck, it refers to the sense of suffocation. In the case of the heart, the metaphorical expressions used to symbolize the pain are manifold: the heart in the grip of anxiety, being shot in the heart, to have heartache, the broken heart or the heart that melts and also the heart beating fast in anger, a heavy and sad heart, etc.

10.2 Treatment of Chronic Disease Through Technical and Nontechnical Skills

This plurality of meanings is well known to those involved in treating people with heart disease, especially when they are chronic conditions. In such contexts, the complexity of meanings represented by the body is redesigned within a unique story, involving one specific person; a specific family background; a specific emotional, social, and economic context; and also a specific group of caring professionals. All these relationships are central to the experience of care. In child care, there is awareness that we are helping trace the footprint that will be distinctive in his/her history and that of his/her family, and this awareness leads us to maintain a constant attention to both the technical and the nontechnical skills [3].

The person and his/her background become central for the medical care. Hjortdahl M et al. [3] have been evaluating the survival, functional outcome, and the quality of life of an increasing population of patients undergoing cardiac surgery and have observed that the physical and psychological recovery of these patients is encouraging, leading to justify the large involvement of hospital resources intended to equip the caring team with the tools necessary to support the pathway to follow. Where there is no cure, but people can live monitoring their health, it is necessary to build an alliance that is sometimes complicated.

Anna's family consists of her parents and a younger sister; she is 12 years old and has a congenital heart defect (hypoplastic left heart) for which she has undergone three palliative surgeries in the first 4 years of life and has always been informed about them. She has regular therapy and is monitored by the same center that diagnosed her condition. She is loved by the doctors and nurses whom she now meets every 3 months, though in the past these visits were more frequent. She is always accompanied by her father and mother, two polite people, who are very grateful for everything that is being done for their daughter. They live in another area, where they have medical references for daily minor issues, but they only trust this center because, they say: "Our daughter was saved here." Recently, in view of the worsening of the clinical picture, the issue of heart transplant has been brought up. When interviewed by the team, the mother said she was worried about her daughter's behavior as she selects foods fearing they might be a source of infection, washes her hands frequently, and is always worried about the wrong dosage of therapy. The cardiologist reminded her about periods in the past where it was easy to make Anna smile and make her cooperative. So by supporting the parents, they have in time become themselves part of an abnormal extended family.

It would be easier if medicine were an exact science never failing to heal patients. The medical team could then even afford the luxury of an intervention with no personal attachment to the patients. In reality, the team needs to establish a relationship with the patient, and especially when dealing with children, it is important to maintain the relationship channel open with the patients and their parents. There is growing evidence that families need help and support. Kendall et al. [4] interviewed 17 parents of children with heart problems at an early stage of non-hospitalization of their children. The drawn conclusion is that the parents need more help and support from health professionals to enable them to manage their children's condition.

Areas of particular interest appear to be those relating to the information parents receive about the health condition, but also the communication between healthcare providers, the guidelines to determine the security levels in the various activities of daily life, and a greater support in dealing with schools. A similar conclusion was drawn again by Kendall et al. in a different section of the study [5], when they interview young heart patients to explore the views of young people with a variety of congenital heart defects, trying to understand what would help them to manage their life better and when and how they would need to be supported.

Here, too, the management of activities and communication about the disease were most often cited as areas that require continuous investment and improvement. This task is more complex when working in pediatrics, where the adult is often the only contact with health professionals. In a research Lee and Kim [6] analyzed the life experiences of ten Korean children and adolescents (14–22 years) with complex

congenital heart disease. The analysis showed that the participants were initially unaware of the true nature of their conditions.

When children start attending school, the physical limitations and overprotectiveness of the parents usually help manifest the real disease condition. In general, healthcare providers tend to provide only parents with information about the disease, and not the patients, who therefore lack knowledge about their disease and often feel isolated and left out. Even Anna, confronted with the difficulty in handling daily life, showed the need to be helped through her obsessive rituals; she is not able to manage the anxiety caused by the worsening of the clinical condition. Her parents, perceiving the issue, ask for help and find the support of the medical team.

In line with what is suggested in the study by Kendall et al. [5], the importance of defining care that can ensure a relationship of continuity in the accompaniment of parents and their children was highlighted. Placing subjectivity as the central theme, the authors point out that the communication of the diagnosis cannot be reduced in a timely and standardized time, such as before the surgery, but it should be seen as a complex process characterized by a continuous monitoring over time and a remodeling of the information in the light of the needs of the patients and their parents.

This study shows the parents' necessity to be able to talk and discuss about the health of their children and to express doubts and uncertainties.

10.3 Parental Involvement in the Treatment of Child's Disease: Taking an Active Role in Emotional Experiences, the Consolidation of Sense of Identity, and Basic Trust

We must not underestimate the fact that for the parents the awareness of the diagnosis is painful, not only in relation to the actual condition of the child, but also in relation to the loss of their image of parents able to protect the life full of hopes they have generated. From clinical experience it emerges that parents often feel responsible (inheritance) for their children's condition and frustrated for not being able to replace them in the experience of illness. In particular, a research by Bevilacqua et al. [7] on 38 couples with children affected by heart disease (from birth to the age of 3 months) has clearly shown how levels of stress and depression were found significantly higher in mothers than in fathers. Parents may therefore benefit from advice given throughout the pregnancy.

The dialogue between practitioners and parents in general appears to be the main guarantee of a communicative process in which the patient and the family become enabled to take an active role and can gradually develop useful skills. This is especially true when the disease is diagnosed in the perinatal phase.

Giovanni, a 5 months old baby, undergoes a surgical catheter placement in the course of treatment of his heart condition. After the surgery, he remains picky and unresponsive for a few days. This causes great concern in the parents until the care workers reassure them about the physiological time required, in light of the clinical event, for the recovery.

The natural process of self-awareness is built through the nurturing mother who provides a sense of what the child perceives, primarily at the body level. Numerous studies show that the child is endowed with competencies to relate with the environment in an active and interactive, mutually "shaping" way. Lichtenberg [8] declared that "during childhood each system contributes to the regulation of the self in interaction with people who take care of the child […]. The activity of each of the systems can be intensified so as to constitute the main aspect of the self" (ibid., p. 8).

There is no such thing as a child separated from his/her relations and the context, making it inevitable to consider being in relationship and the social dimension as constitutive of the person. The body keeps traces of the world it lives in [9]. In line with this, Liotti [10] reminds us that "the brain and memory pass into the body, they are in the body. The self and consciousness are inseparable from the body." Winnicott [11] talked about handling and empathic holding.

All this is well established in terms of physiology, but where the body signals its fragility, things get complicated. Treatments must take into account the significance of the relationships, primarily with the parents. The importance of contact, its quality, its context, the ways, the intensity, etc. in pathological condition is therefore called into question.

For some time it has been shown [12–16] that the factors that allow parents to establish a public emotional attunement and an ability to respond sensitively and accurately in relation to the needs of proximity, contact, and protection of the child are in direct correlation with the mental representation of the child in the mother, as the mother projects to the child's both perceptions and experiences.

As already mentioned, when the fetus and newborn shows a pathology, the confidence in their ability as parents is questioned, by the mother in particular, to generate a healthy child, as well as the confidence to be the first guarantor of its survival. This can interfere with the basic trust of the parent with respect to their ability to emotional containment [17], as well as the consolidation of what is the basis of the child's sense of identity.

In the hospital setting it is important to remind mothers that in order to build confidence in their children what is necessary is not the absence of disease, but an ideal combination of sensitivity to the individual needs of the child and of the self.

Carla is 20 years old, like her partner. She is the mother of Alessandra, a 4-month-old girl who has never been out of the neonatal pathology ward where she is hospitalized for a congenital heart defect. Her partner asks for psychological support for Carla as he notices that she looks sad and feels emotionally distant. Carla describes how the nurses, in her opinion, consider her too young and consequently inadequate as a mother, "even when I have to give the bottle and Alessandra cries the nurse takes it from me and does it herself … the same to change the diaper. If she cries at night I'm afraid to disturb. I do not know how I can manage at home …"

In such a situation, health is influenced by the mother's attempt to regain self-confidence toward her womb, first, and then the baby, and to take over the

relationship with the baby, together with the father. Health professionals necessarily step into that mother-child relationship and family unit. They take on the dual task of being both guarantors of the survival subjects and reinterpreting the conduct of the infant and child, by discriminating the pathological from the physiological. It is crucial to foster the development of interaction between mother and child to avoid the risk of operators becoming a substitute for parental roles. The operators often become key elements for parents to get in touch with the children and decode their needs. Sometimes parents feel unable to deal with their children's body and leave the specialists with this responsibility.

The team of specialists provide those points of view that, in pathological conditions, seem to be necessary to complement, support, and, at times, correct and reread the signals coming from the baby. The multidisciplinary team, before the birth of a child with complex disease, needs to support parents in the caring tasks. Probably, in the case of Carla, the young age of the couple leads, more than in other situations, to a replacement of the roles. The parents' effort to achieve confidence in their skills can also affect the child. The parent tends to see the children as scarcely able to understand the signals of their body. The awareness of one's body is questioned to the point that there is a failure in decoding a child's psychic communication about their own health, which has to do with the unity of mind and body.

In all these cases, offering support to the mother or the parents necessarily involves empathy, which is, by definition, sharing and experiencing the feelings of a certain person, even if only temporarily [18]. Being empathetic, in this specific context, means understanding the fragile condition of a young couple with a child who has been hospitalized since birth. This is important, as long as it does not result in being overwhelmed by the intensity of feelings or taking responsibility for the situation. The dual task that healthcare people face is therefore to capture the experience of others, recognize it, and at the same time be aware of their own feelings and keep the two separate.

10.4 Supporting Parents in Taking Care of Their Child with a Heart Condition

Luca tells his parents about a recurring dream of being bitten in the abdomen by a roommate in the hospital where he was admitted for a heart condition for which a pacemaker was required. The parents ask the cardiologist if it is possible to talk to a psychologist as they feel concerned by the dream and also by other behaviors, such as little interest in playing games, especially games involving physical contact. The subsequent pacemaker infection and the replacement of it allows Luca to resume everyday life, and at the same time the parents and the team recontextualize the data as the attempt of the child, with the resources available in a 4-years-old boy, to communicate his needs.

The patients, however young, are the main protagonists of the condition and they should be listened to, as they can tell important information about their bodies.

A person who is confronted with a serious heart condition often has a long history of investigation, analysis, aggravations, and improvements related to that operation or other therapy, in a life where the hospital, be it near or far, becomes a familiar place. Supporting parents in understanding what their children say is an important part of the treatment.

Salvatore, who has a diagnosis of hypoplastic left heart, lives in southern Italy and comes to a hospital in the north for the regular inspection required by the treatment plan. Caregivers are alarmed by what they see as a serious negligence of the family that underestimates the clinical picture, as they arrive with adequate medicine for only 1 day, not considering the possibility of unexpected, prolonging absence from home, which could endanger the child's life. The mother is focused on a marginal aspect, according to the medical staff, which regards to the feeding of the child, and always asks to consult a pediatrician with specific expertise on nutrition. This creates a stall condition and misunderstanding. Empathy falters, and the recognition of the mutual positioning between medical staff and the mother is poor. This situation improves when the staff finally decides to accept her request without sacrificing the explanation of the clinical side.

In this case, the objectives of care focus on the complex task of integrating the needs of the child and family with the clinical requirements. In line with that, among nontechnical skills, operators are required to have the following abilities: emotional stability, to learn from any experience of exchange and interaction, to refrain from acting out, to take responsibility, and to act in a manner understandable to the others so that they can consciously participate in the treatment.

In the case of Salvatore, understanding the maternal perspective made it possible for the team to define the right path to follow and reinforce the therapeutic alliance. The nontechnical skills, interweaving with the technical skills, have allowed a better therapy, as everybody's needs were taken into account.

Michele is a 16-year-old patient who has undergone a heart transplant. He belongs to a family perceived by the medical team as simple and not always completely adequate. The boy, at this stage of his life, is often showing a challenging attitude. Blood tests begin to indicate the presence of complications, and the team suspects possible light drug usage. The parents are upset both for the blood results and for the suspicions, as they reassured the doctors about the drugs taking. Michele said he can manage to take medications and chooses to have a hostile attitude both with the family and the doctors.

Understanding the complexity of the situation and the risk of damaging the alliance with the boy and his family, the care team decides to reread the dynamics involved and reconsider the course of care, after listening to the family's requests. It is the interaction between the technical and nontechnical skills which allows the team, not without little effort, to explain to Michele and his family the clinical situation and the actual state of health.

Much has been said about the relation skills of the team, particularly the ability to communicate, and its effect on the overall health of the patient and his/her family. Referring to Green [19], it was proven that the better the mother's understanding of the health conditions of their newborn children, the greater also the children's safety. Also, in a previous study, O'Brien et al. [20] reiterated the need to provide parents with the knowledge and skills for the care of their child during the period of hospitalization, emphasizing that this requires the joint efforts of a multidisciplinary team in providing clear, concise, and consistent information.

McCusker et al. [21] conducted a study on 90 children (aged 4–5) where, after the baseline evaluation, families were assigned to an intervention group or a control group. The usefulness of the intervention was assessed by analyzing the maternal mental health and the dynamics within the family. The research showed that the children in the intervention group were perceived as "sick" less often from their mothers and they also missed fewer days at school. This study also emphasized the reduction in maternal concern about the child's behaviors up to a year away. Similar considerations have also emerged from a study by Montis et al. [22], aimed at better understanding the influence of communication processes of psychological interventions and support for parents of children with heart disease.

The study compared the anxiety levels of 380 parents of children already used to be treated in hospitals, with those of 240 parents of children at their first access to the structure. The authors found that appropriate interventions, in particular an effective communication in an appropriate environment, reduce the anxiety of parents waiting for medical evaluation.

It also showed a direct impact on the child, whose psychological well-being was directly related to the emotional state of the parents. Other research explores the importance of legitimizing the questions that parents ask themselves about daily life in relation to the child with the disease. The research shows that parents sometimes perceive the medical staff as "too busy" to respond or they fear their questions are not relevant [4]. Some of these questions relate to the need for normality and the need for protection, which depend on multiple factors within the child and the family and also by external factors. Rempell et al. [23] show that parents often hold in solitary confinement, even within their own home, the young patients during the waiting period of the intervention.

In addition to this, there is a need for protection with respect to other parameters that are the subject of much attention by the family and by the medical team. O'Brien et al. [20] focus the attention on the fact that school attendance by children

with congenital heart disease involves elements of concern for their health and safety. In this study, after an overview of the health problems that involve school-age children with ConHD, it is argued that the healthcare needs of the child require an extended work of individualized educational planning, involving both the health field and the school.

10.5 Supporting the Patient in Taking an Active Role in His/Her Therapeutic Procedure Within a Process of Dynamic Change

To add complexity, operators working in the pediatric field are well aware of how the information needs and attention in the course of treatment are extremely sensitive to age and role. The child, in fact, at different stages of his/her life has different skills and needs, very often related to the procedures they have to deal with (such as the injections), while parents focus their attention on the diagnosis and the prognosis and also, empathically, they feel their child's fears. As the child grows and starts thinking for himself/herself more, he/she is faced with the presence of the desease as an additional developmental task, which will accompany him/her during adulthood. Anna, a teenager, should and would like to deal with a changing body, but at the same time is measured with a worsening condition of fragility due to the presence, and in her case the worsening, of the disease. Whereas her friends are busy experimenting with the building of adolescent identity with the physiological fear of the new, Anna, as often happens to children suffering from severe and chronic organic disease, is facing fears amplified by the presence of the disease which imposes limits, boundaries, and control and brings with it a sense of insecurity.

It is well known that teenagers are, by definition, dealing with developmental tasks that have to do with a renegotiation of the relationship with the adult, with the limitations and the reformulation of their identity. Tong and Kools [24] show that parents of adolescents are called to move gradually from having total control over the health of their children to a shared management of these aspects, however expert they have become over the years about the disease and its management.

The disease is present in a particular time, when the teenagers try to break away from parental authority, although the responsibility for the management of the disease stays with the parental figures who are "guarantor of the safety" of their kids. Medical treatment should make teenagers more responsible for themselves, in order to create a valid connection with adulthood. Anna's story, which indicates the presence of a large reliance on medical staff, shows a situation that could be called, from the point of view of communication, transparent. Anna has always known everything about her condition, and the medical staff informed her of the possibility of a transplant when they thought it might be necessary.

This becomes particularly important in adolescence when kids can struggle to perceive themselves as their peers because of frequent hospitalizations, physical limitations, and concerns about their health, as these aspects can largely affect their social relationships. In adolescence the treatment has to do with the repositioning of

the teenagers within the therapeutic relationship, and it is necessary to take into account their individual evolutionary stage. It is therefore important to gradually develop a communicative exchange between practitioners and patients, alongside the already established one between practitioners and parents.

In the article mentioned above, Tong and Kools [24] talk about the discomfort experienced by teenagers when not being heard and "taken seriously" by the caregivers. The best way to counter this experience is the ability to communicate. When Anna is informed about the heart transplant, she loses confidence in herself, in her ability to distinguish what is good and what is not. The transplant seems to elicit in her an emotional disorientation. To lose such a crucial organ and receive a new one from an unknown donor is obviously a complex issue to deal with. In his book entitled *The Intruder,* Jean-Luc Nancy says that the patient who has received a new heart never accepts it; the process of assimilation of a foreign organ is impossible. It will always be an intruder. It is important to understand what the therapeutic proposal advanced by practitioners means for Anna, and what can be done by the team to best face this experience.

If you want to develop meaningful interventions, especially during times of change such as adolescence, the perspective of the patient and the family must be understood and included. The assessment and recognition of the evolutionary changes that occur for both adolescents and their families and the impact of chronic disease on these changes are essential to support the path to health. Stereotypes should be recognized and changed in view of the uniqueness of the patients being treated, the changing roles of those around them, the reorganization of responsibilities, and the recognition of losses related to the disease condition. The whole team has a key role in supporting patients and their families favoring the identification and use of resources to make sure that, in this delicate transition phase, chronic illness is not completely ignored or, at the other extreme, regarded as the central aspect of identity.

Albert et al. [25] carried out a study on 38 young heart transplant patients in early childhood or in adolescence, who were at the time of research between the ages of 16 and 34 years. It was evident that, compared to a group of healthy peers, they showed a significantly reduced quality of life in many psychological and somatic domains. Almost 50 % of patients reported this situation. These results have highlighted that with young patients struggling in the transaction to adulthood, healthcare providers need to be particularly attentive to the psychosocial dimensions, in order to prevent certain behaviors that can compromise the quality of life in the long term.

The feelings of "invincibility," concentrating on the here and now, the nonconsideration of the outcomes and consequences of future actions, are considered as specific risk factors, which are typical in the transition from childhood to adulthood, for an adequate management of the care pathway. Clinical practice has in fact over time underlined how adherence to treatment succeeds better if there is the active involvement of the teenagers in their therapeutic procedure, starting from the need to guarantee a basic understanding of their condition and overall clinical situation. Bar-Mor et al. [26] examined the cognitive processes that influence the involvement

in physical activity among 100 adolescents, 55 males and 45 females, aged 12–18 years, with mild or moderate forms of congenital heart disease, reaching two very interesting conclusions.

The first was that, in this group of people, beliefs regarding one's own self-efficacy, rather than the severity of the disease, were the most influential factors in determining the conduct of adolescents. The second is the importance of the role played by the recommendations of the cardiologist in the determination of parental attitudes and perception of self-efficacy of young patients.

It is evident that the care team cannot but engage in a relationship with the patients, not only for those aspects of therapeutic adjustment always present in a chronic illness, but also of the change that comes when the patient is confronted and cannot find an answer in the parents. Parents are asked to understand the clinical picture, adhere to the therapeutic proposal, and support the child and his/her health.

10.6 Internal Relations and Emotional Process of the Caring Team

But the dynamics of care also affect the internal relations of the caring team. Just like in a family, there are different positions, so the individuals within the team are carriers of views, thoughts, and considerations which, fortunately, are not always the same. The more complex the situation is, the more complex are the treatment choices. Sometimes the relational dynamics are manifold and complex, partly because the clinical opinion, the reading of the symptomatic aspects, and therapeutic indications may differ significantly. In most cases, the medical staff, confronted with the emotional surges and clinical comparisons, reach a shared proposal to submit to the family. The team provides the family with a protected space. This is the well-known complex web of experiences and emotional intensity that distinguishes the doctor-patient relationship.

It starts with the diagnostic communication and goes on as a complex process of mediation that takes into account many parameters, including the ambivalence, a mixture of gratitude, and the need of compensation that parents experience against those perceived as more competent and adequate in the care of their children. The operators often feel they are projected into the role of hero and, shortly after, deeply devalued.

In ancient Greece, there was a ritual: two people, a man and a woman, each year were consecrated to play a dual role. Critical to the welfare of the group before, and scapegoats afterward, they shifted from being fulfilled in their desires for a year before being symbolically loaded for all the ills of the community and expelled because they were considered cursed. They were called *pharmakoi*. The name, which now coincides with a chemical substance administered to the patient, however, was once primarily a natural person. In Athens, then, when the evil inside had been expelled from the community and transferred to the two *pharmakoi*, these were brought out from the confines of the town (polis) to be burned and their ashes scattered to the wind, so the accumulated evil of the polis would be completely healed.

Some claim that this is the root of burnout: healthcare workers are always exposed to the transference of the suffering of their patients they are afraid to absorb. Patients are respectful, almost reverent, bring gifts, and show gratitude and affection as long as the physician does not become their scapegoat and, as such, responsible for everything that does not work. The team that takes care of heart disease is even more vulnerable to this relational dynamic because it is a specialty niche, and medical centers for heart diseases are only a few and of excellent standard. In the pediatric setting, as already argued, there are also the intertwined themes of addiction and parenting. The team may, themselves, assume parental roles in contraposition to the parents.

10.7 A Possible Way Forward

In order to understand and build a therapeutic emphasis nowadays, we talk about narrative medicine. Even William Osler, a Canadian humanist physician of the early twentieth century, said "it is much more important to know which type of patient has a disease than to know what disease a patient has." Perhaps we can replace the word "sick" with the word "person," but we agree with his statement. When the medical team welcomed Anna who was still in the womb, they became aware of her parents' background, and they made advanced diagnostic hypotheses and devised a plan for clinical tests in order to understand what was happening in the little patient. The team did all this aware that, after this stage, their diagnostic information would be shared outside the narrow circuit of experts. There is always a need to know further elements, not only relating to the diagnosis, which should become an integral part of the story of a person, in his/her family, and their relationship with caregivers.

In conclusion, we could mention Buzzati's "Seven Stories" where the main character, Mr. Corte, is left to himself and to his fear of dying; thus, he begins a metaphorical descent through the 7 stories of the hospital in which he is admitted and starts to be thought away "from the world of normal people." The accompaniment offered to our young patients and their families will not let them feel alone and ungrounded, because, as we read in Buzzati's story, the fear of the disease is much more unmanageable when it is interwoven with the fear of the unknown and the unspeakable.

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Art Therapy as a Psychological Intervention for Hospitalized Congenital Heart Disease Children

11

Emilia Quadri, Edward Callus, Elena Palmero, and Cristina Farè

The use of art as a form of therapy is based on the idea that artistic images can help us understand who we are, to express feelings and ideas which are impossible to communicate, and to enrich our lives through self-expression. The efficacy of art therapy in the hospital settings has been confirmed for many different pathologies. It has been seen that the therapeutic programs of artistic activity in hospitals bring clear psychological and also physical benefits. A key aspect of art therapy is empowerment: through it there is the capacity to improve the self-care and the handling of oneself of the ill people. Since 2009 we have applied the methodology of art therapy as a psychological support intervention for the children hospitalized in the Pediatric and Adult Congenital Heart Disease Centre at the IRCCS Policlinico San Donato University Hospital in Milan, Italy. This chapter is a description of art therapy and its implementation for congenital heart disease patients.

In recent years, the benefits of art have been rediscovered; art can indeed be the means to allow us to overcome adverse circumstances or life problems, recover from traumatic experiences or losses, relieve pain, and give meaning to what happens to us [1]. The use of art as a form of therapy is based on the idea that artistic images can help us understand who we are, to express feelings and ideas which are impossible to communicate, and to enrich our lives through self-expression [1]. The term "art therapy" refers to a form of relationship in which the parties, in some way, are inverted, since it is the subject to work actively – while the clinician remains in the background – to get in touch with their essence and their "inner experience" [1].

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The use of this therapeutic technique in hospitals has recently had several confermations, following practices which are more or less structured and in relation to different pathologies [2–4]. In these experiences, the substantial effectiveness of art therapy in its various forms has been shown. In these contexts art therapy is often successful in leading to achieve a self-awareness of the disease and the discovery of the implications that the disease has on a daily basis, which would be difficult to achieve through words alone [1]. Finally, it is an alternative channel of communication that is particularly useful and effective in the circumstances in which a particular emotional state, discomfort, or fear cannot be expressed through words, as often happens with children and adolescents [5].

The application of art therapy techniques in hospitals has also been favored by the fact that it has been recognized in 1994 as a "mind-body intervention" by the National Center for Complementary and Alternative Medicine, taking into consideration the effectiveness that the expression of the self through the creative process has in positively influencing the body. It has been seen that the therapeutic programs of artistic activity in hospitals bring clear psychological and also physical benefits: from stress, anxiety, and depression reduction; an improvement in the ability to communicate symptoms; a reduction in negative symptoms and an increase in energy levels; improvements in blood pressure levels, heart rate, and respiration; and the capacity to assess one's own general health [6–8].

Artistic expression can then be very useful in assisting children and adolescents who have to prepare for medical procedures such as surgery, injections, and intravenous catheterization, especially when combined with play activities. In drawing, as well as in all forms of artistic expression, the child and the adolescent represent their feelings, emotions, and experiences; every little detail in a drawing can tell us about their inner world bringing private and non-tangible aspects that are personal and specific to the person, more than in a clinical diagnosis. During hospitalization there is often a state of crisis due the trauma of the disease, and the assessment of the patient's psychological state through the artistic activity is a useful tool to allow the young patients to express their feelings. At the same time, this kind of activity provides the therapist and the physician the information necessary to understand the level of psychosocial functioning, the symptoms, and perceptions about the medical procedures and hospitalization of these patients [5].

Art therapy can be considered as the combination of two disciplines, art and psychology. In its definition, the aspects of the creative process of human development, behavior, personality, and mental health are also involved.

It was created from the idea of using the means of expression of the visual arts to create a new opportunity and communicative potential, fostering the ability to properly regulate emotions, facilitating their elaboration and also their reproduction, in a certain way.

The Greek word *therapèia* means to "pay attention," a meaning that emphasizes the profound meaning of art therapy from two points of view. Firstly, there is the specialist who follows the subject while the work is being produced; he/she may be a suitably trained artist or a psychologist. Their leadership is important in the therapeutic process, as the care and support provided by this relationship are essential in order to able address the artistic experience and help the individual to discover the

meaning connected to it. The second important aspect is the attention that the person focuses on his/her activities, to give sense to the artistic product, that is finding a story, a description, or meaning which adequately defines it. In this sense, few other art forms depend, as this does, on the active participation of the subject.

The therapeutic function of art is therefore found in between these two definitions which try to give a specific meaning to the experience of art therapy. The first one calls into question the conviction that the creative process in itself has a curative power. This conception implies the idea that making art is a therapeutic process; it is seen as an occasion to express oneself in an authentic way. This experience, with time, can lead to the realization of oneself, to the ability to take care of emotional wounds and the capacity for transformation. The second type of definition is based on the idea that art is a symbolic means of communication: such an approach which is often referred to as "psychotherapy through art" outlines the value of the artistic products as a means of communicating problems, conflicts, and emotions. The artistic image acquires a meaning since it encourages the exchange between therapist and client who is more apt to a better understanding of himself/herself. With the guidance and the support of the art therapist, art promotes the awareness of one's situation, helping to resolve conflicts and to formulate new perceptions that can also lead to positive change, personal growth, and an increased physical and mental health [1].

The *instruments* of art therapy, which aim on the fluidity of play and imagination during the therapeutic process, can offer a specific contribution to the subject enabling them to disengage from the degree of rigidity of their mental schemes. It is often even more significant on the level of dysfunction of the subject. Again, in art therapy, a paper can become a symbol of one's own psychic space – an important assumption in psychic growth which was already mentioned and which corresponds to the states of mind, thoughts, emotions, hope, and potential of the individual: the "Finite Self which is surrounded by the infinite world" [9].

The utilization of art in the treatment of psychological and psychosomatic aspects in illness became widespread for various reasons but also because through art there is the possibility to transmit important messages, which are both conscious and unconscious, both to the psyche and also to the body.

Some studies have investigated the effectiveness of art therapy on psychological symptoms, whereas others have focused on the reduction of the physical symptoms and some others on the importance of the experience of the participants. A key aspect of art therapy is empowerment: through it there is the capacity to improve the self-care and the handling of oneself of the ill people [10].

Significant improvements were reported in general when it comes to depressive symptoms, stress, quality of life, global health, motivation, the ability to talk about the physical and mental health, and coping resources, whether the sessions were conducted in groups or individually, even in the absence of a real improvement in physical health [11, 12].

Another therapeutic aspect in the use of art therapy in the hospital setting observed in the various studies is the strengthening of the self of the sick person. Through a qualitative analysis, it was found in the study by Oster et al. that art therapy can be a valuable tool for personal growth, being used by patients to strengthen their boundaries

in relation to others' needs – which are often not respected, increasing anxiety in their already difficult situation – and also to understand their condition [13].

In addition Collie et al. observed how art therapy in oncology is useful in enabling the patients to see their experience in a more clear manner and to stimulate change also on a behavioral level. In this way the patients manage to maintain a stable sense of self, reinforcing or amplifying their perspectives and experiences [14].

The analysis of the art therapy therapeutic process carried out by Gabriel et al. has outlined different types of change in the patients: during the various types of art therapy sessions, the patients can shift from a passive mood to a more active one, from a state of stress to a calm one, from a feeling of fragmentation to a sense of unity, and also from a state of psychological isolation to a greater interaction with the other participants in the art therapy groups [15].

Finally, in the analysis of the interviews carried out by Gotze et al., some significant dimensions of the utilization of art therapy have emerged: the emotional stabilization, a deeper and increased expression and personal growth, improvements in tackling the disease, and a more appropriate communicative capacity [16].

The perception of the effectiveness of art therapy in the medical context has been investigated in a study by Forzoni Perez et al. carried out in 2009 at the Oncology Day Hospital in Siena, Italy [3], in which patients were waiting to be subjected to chemotherapy. The patients had participated in some individual art therapy sessions (the average was 4–5 sessions), during which they were asked to create a collage composed of images relevant to them and then to give a title to their work. They were subsequently asked if they thought that the art therapy intervention had been helpful and in what way.

Only 5.5 % of the patients believed that the intervention was not effective – probably because they had participated to a single session – considering the methodology childish and superficial.

On the other hand, 94.5 % of patients reported the positive effects of the intervention. It became clear that there is a continuum of experiences that the creative process offers the individual: a lighter and often sensory experience (e.g., "I was able to relax") to a deeper experience (e.g., " it helped me to get in touch with my unconscious"); the attention is no longer directed only to the interaction between patient and therapist, but it becomes a triadic relationship: patient-therapist-image. The patients affirm of how their artistic products help them to better understand themselves ("I was able to talk to myself, as if my true self had gone out from me, and it was projected on the sheet of paper, and was talking to me … I could see myself on that page").

Art can become, especially for the child, an essential balancing element between the intellect and emotions. It is indeed an activity to turn to when something, even unconsciously, is disturbing, especially when words are not sufficient and adequate. Through drawing children can talk about themselves and their world by sending real messages, thus learning new ways to release emotional tension.

The professionals involved in the care of the physical and psychological health of hospitalized children and young people agree that the experience of illness and hospitalization that requires a separation from parents can have a profound influence on the development of the patients and their emotional growth due to the loss

of independence and control, the fear of being continually subjected to medical treatments and surgeries, and the constant fear of death. Therefore, the psychological needs of young patients are the focus in the use of art as a form of therapy in this context [5].

In the drawing, as well as in all forms of artistic expression, the child and the adolescent represent their feelings, emotions, and experiences; every little detail in a drawing can give insight about their inner world giving relevance to private and non-tangible aspects that are personal and specific to the person who is always more than just a clinical diagnosis.

Another important function of art therapy with children and adolescents suffering from some illness is in its use for the understanding of how they perceive and conceptualize their body image and identify their physical symptoms. The body image is the mental representation of how individuals perceive their body when it comes to the physical, emotional, and interpersonal aspects. The integrity of the development of body image is challenged by the appearance of the disease, pain, symptoms, and the surgical procedures it entails. The use of a simple body scheme given to the patient to complete can help health-care professionals to understand the areas of pain perceived by the child – which is not always possible to communicate verbally – in order to understand and better deal with the symptoms and allow the expression of their perception of the disease and the way they see it affects their body, especially when it comes to adolescents [5].

Due to its specific characteristics in this context, art therapy takes on a therapeutic value: through its functions art, as has already been observed, acts as a container by regulating the most painful emotions that may emerge without provoking dramas, turning them, through the artistic medium, a potentially traumatizing experience into a creative adventure, moving in a positive way the concentration toward play and creativity, taking it away, at least for some time, from a place filled with anxiety, that is, the hospital and all that it entails because of the disease.

The child and adolescent patients need to feel that they can take control of their disease, that they can transform it, comment on it, and represent it, even if it means bringing out, for example, gloomy and depressive thoughts, which are congruent to the anxieties caused by disease in the children and their families [17].

These young patients use drawing and artistic expression, regardless of their specific content, to reclaim what happened to them and to become familiar with the disease and with their fears. The experience of being able to have control on their feelings and their sensations, even if on a sheet of drawing paper, is very important given that they cannot control their medical condition.

Art therapy is one of the few therapeutic activities in which the individual becomes actively involved in treatment through the creation of a tangible product – which serves not only as a form of communication of feelings and experiences but also as an external and visible documentation of one's self. For the children admitted to hospital, the active quality of the making, drawing, cutting, pasting, and producing relieves feelings of helplessness and lack of control associated with physical weakness and hospitalization [5]; the ability to reverse this feeling of passive powerlessness into feeling creative and active is one of the most obvious benefits of art therapy [18].

Since 2009 we have applied the methodology of art therapy as a psychological support intervention for the children hospitalized in the Pediatric and Adult Congenital Heart Disease Centre at the IRCCS Policlinico San Donato University Hospital in Milan, Italy. Currently, this hospital is one of the leading international centers for the treatment of congenital and acquired heart disease. The workshop has been structured according to a model that takes into account the specific needs of cardiac patients and is run by two psychologists responsible for the project, an artist and a peer counselor, assisted by several trainee psychologists and psychotherapists.

The intervention was organized in a group setting with a frequency of three times a week in a large room in the ward. The workshop was designed to be applied to three specific moments of hospital admission: the preoperative phase, the postoperative phase, and during emergencies. The activities are scheduled and programmed depending on the time of hospitalization, the patient's age, and clinical condition.

Therefore, the opportunity to gain experience with peers who share the same fate of disease is offered and every artistic production is shared within the group. All proposed activities are aimed at the expression of the feelings of the patients with respect to the physical and mental suffering related to their heart; in fact it has a high metaphorical value, as a vital organ and place of emotions. Family members and patients hardly talk about it, both in the preoperative phase and post-intervention, also with the appearance of the indelible signs of the disease, e.g., the scars. It is therefore of great value to offer them the opportunity to discuss, give vent to their fears, and understand that they are not alone in facing this difficult journey. The main activities have been conceptualized taking into account the most pertinent literature mentioned previously, and they are presented here.

11.1 Presentation of the Self

Participants browse through magazines available and choose pictures they consider representative of their current condition.

Age: From 6 years.

Task: Participants browse through magazines available and crop images that are interesting to them for several reasons. It is then proposed to create a composition that makes sense to them and to give a title to this composition.

11.2 The Scribble

It is used with the children in order to allow them to express feelings and fears, exploiting their propensity to creativity.

Age: From 7 years onwards.

Task: Participants rest a pencil on a paper (closing their eyes and using the non-dominant hand, if possible). They are then asked to follow the movement and rhythm of their breath with the pencil, until they feel the desire to stop. When

looking at the scribble that appears on the page, the lines may suggest an image, which can be completed with colored crayons. The picture which forms the doodle is well suited to become the inspiration for a story or a fairy tale.

11.3 Body Schema

Participants are asked to complete a model of body schema given to them, in order to see how they represent themselves and their pathology.

Age: From 5 to 14 years.

Objectives: To assess the perceptions of individuals about the intensity of the symptoms, the "place of pain," and if it has a definite shape or color.

Task: The patients are asked to close their eyes and, starting from their feet, to note any trace of feeling or pain they experience in every part of the body. They are then asked to fill in the scheme with lines colors and shapes, without worrying about the realism of the final drawing. Finally, the task is completed with the addition of some explanatory sentence.

11.4 Tree Test

Patients have the task of designing a tree freely, which is then brought to the attention of psychologists, who evaluate the structure of personality based on the current age of the patients.

Ages: 4–5 years old onwards.

Objective: To be able to understand the most authentic but hidden aspects of their personality. The tree is the symbol of the self, that is, the energy that involves the whole person, and it can reveal one's true essence, representing the character, emotions, and the own unique emerging personality. Every detail of this graphical test has a specific meaning that is transformed into plain language: the positioning of the tree on the paper, the roots, the trunk, the leaves, and the presence of other elements.

Task: It is better to ask the subject to perform the drawing "free hand" without the help of rulers that hinder spontaneity in the execution. The following instructions are given: "Draw a tree, whichever tree you like as it comes to your mind. After you complete it you can then color it if you like."

11.5 The Drawing of the Self (Pre- and Post-intervention)

Subjects are asked to represent themselves freely – or their heart – before and after the intervention, in order to compare the two images and to assess the effects of the surgery and whether and how the psychological support influenced the emotional distress.

Ages: 9 and older.

Objective: To assist children and young people and give them the opportunity to manage their feelings of anxiety and fear before and after the interventions.

Task: The patients are asked to represent their illness themselves – or their heart – through the "art" by using all the available materials. The therapist then explores the content of the drawing or artistic creation with the patient, helping them to clarify feelings and perceptions and to identify the people in their life who can support them.

11.6 Activities with Medical Material

Age: From 3 to 7 years.

Objective: To promote a sense of competence and self-efficacy, leading to familiarization with the medical material, an inevitable part of their everyday life in the hospital.

Task: The therapist reads the children a passage on the specific topic of heart disease, medical treatments that should be followed, and the most common sensations that ensue; after that the children are asked questions about their experiences and their feelings in order to see if they are similar or if they had a different experience. Finally, they are asked to "decorate" their cardboard bear with the medical material available ("Now you are the doctors!") and to draw on its face any expression deem appropriate ("How does this teddy bear feel?").

These techniques which have been illustrated and which are used in our laboratory have the common goal of allowing patients to express themselves freely and in a protected environment, and they have been selected specifically for them in order not to create any constraints with respect to the tasks suggested; the priority is the possibility to express one's feelings freely.

11.7 Case Report: Giuseppe, 13 Years

At birth, Giuseppe was diagnosed with a complex heart disease: mitral atresia, single ventricle, and coarctation of the aorta. He had to undergo cardiac surgery before the age of 10 days and subsequently at 2 years and 5 years of age. In April 2012, during a game of volleyball, the patient experienced abnormal cardiac symptoms and he had to be hospitalized. During this hospitalization, which lasted several weeks in June 2012, a defibrillator was implanted and Giuseppe had the opportunity to participate in various art therapy activities.

The contact between Giuseppe and the service offered by the art therapists is mediated by his mother, who signals to the nurses that her son is very anxious.

In fact, when Giuseppe is first approached by the art therapy staff and the laboratory is presented to him, he is particularly anxious, but he still agrees to participate in the activities the next day, even though initially he is not particularly enthusiastic about it. As already noted during hospitalization, the boy appears quite anxious

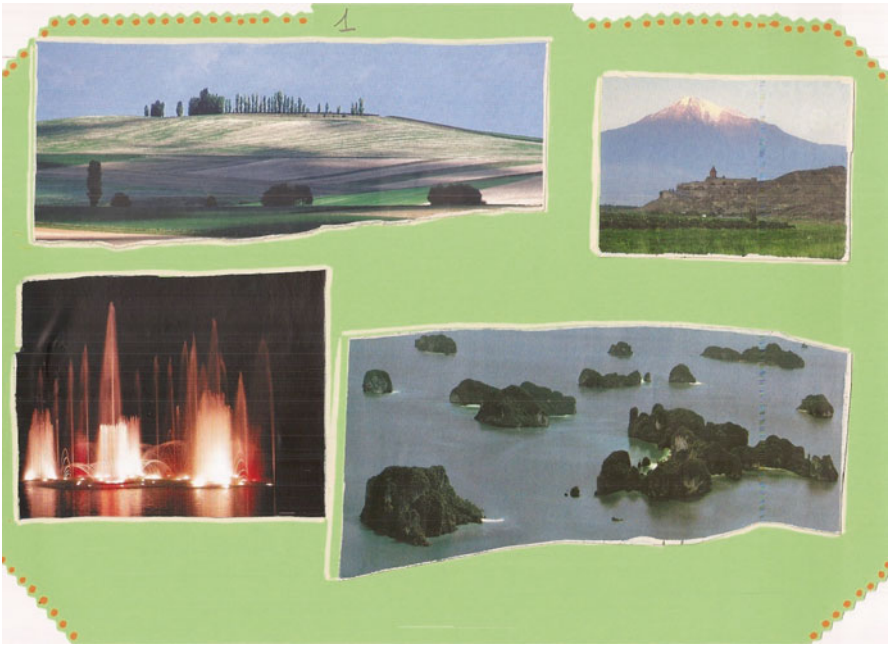

Fig. 11.1 Giuseppe's first Collage

(anxiety expressed by nonverbal behaviors such as sweating, shaking, and the expression of continuous fear of dying) and is particularly scared about the possibility of having to undergo another surgery.

Initially, the patient has difficulty in relating to other people; after several days the boy decides to participate in the activity of art therapy, despite his apparent inability to adapt to the proposed activities in the laboratory.

Giuseppe was contacted several times, and finally, he develops enough confidence to perform certain tasks, including the collage for the presentation of himself; also by observing the work of others and getting some ideas from them, he manages to complete his collages: he pastes images depicting landscapes that seem to not to make any sense, and when he is asked to explain them, he does not provide any information about it (Fig. 11.1). The same attitude is maintained in the second collage that he makes (Fig. 11.2).

The next day, he adheres to the proposal to "construct the heart": the attitude of shyness persists; he speaks only if he is asked some questions but he still proceeds with the activity. He slowly begins to tell us about his creation and he finally manages to confide that it is an especially difficult hospital stay for him and that he cannot wait to go home.

In particular, he says he is struggling, mainly because of the lack of friends and classmates and because he is afraid. After about a week of hospitalization, it is proposed to Giuseppe to draw a tree in an individual session in his room. During

Fig. 11.2 Giuseppe's second Collage

this activity, he relates his experience of a previous hospitalization occurred in a hospital in Potenza, in which he only remembers an unpleasant environment and a place full of older people. When he finishes the drawing, he observes it and says that it seems to be made by a 5-year-old child; he believes it is an ugly drawing: the therapists reassure him on the subjectivity of the beautiful and the ugly and remind him that there is no assessment with respect to how beautiful his drawing is (Fig. 11.3).

After a couple of days, he is eager to participate to the laboratory, and a stronger and more active membership emerges. After 12 days of hospitalization, it is communicated to the family that the boy will have to undergo surgery, and the mother is very worried because she thinks that the child will not be able to have a normal life like that of his companions. This news takes its toll also on Giuseppe, and the psychologists help him to prepare to the intervention, by accompanying him do a preliminary round of the ICU in order to enable him to become familiar with this environment and especially to emphasize the characteristics that he finds interesting – nice and welcoming staff and a place that "looks like a spaceship." The intervention has a good outcome and Giuseppe is back in the department after a few days.

During the postoperative phase, the boy complains about physical pain and difficulty in relating to parents, whom he finds overbearing. He tells us his fantasies and his plans for the future: he would like to finish high school and then stop and rest for a few years, to later embark on a submarine on which he would sail "away from everyone."

Fig. 11.3 Giuseppe's Tree Test

From this fantasy, he develops a story, which he entitles "The submarine which lived twice," which he would like to be read to the art therapy team.

The next time he participates to the laboratory, he arrives accompanied by his parents and he is smiling; he already knows what activities he would like to participate to.

Giuseppe's course ends on a positive note in his participation to an activity including cardboards proposed by the artist of the department (Fig. 11.4): the choice of colors and the progressive increase in adherence to the activities and enthusiasm lead one to think that the boy has now learned to express his emotions more freely.

Giuseppe is discharged the following day: he returns after a month for a checkup and everything progresses well; although he still feels a bit weak, he seems more calm. His case is a clear example of how art therapy can have a positive influence, as it allowed him to socialize with others, to share with them what he feels, and also to deal with the hospitalization and intervention with an increased awareness and tranquility.

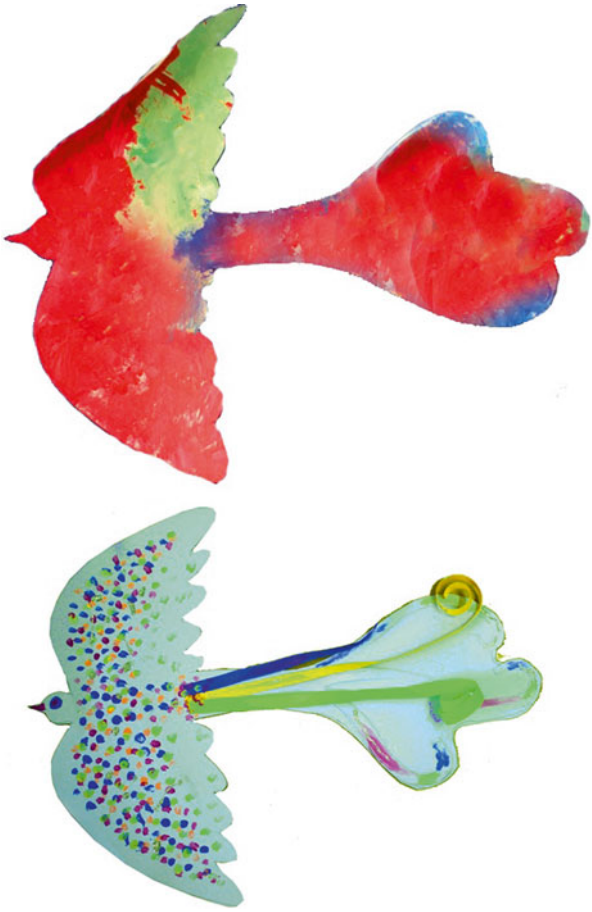

Fig. 11.4 Giuseppe's Cardboard Birds

11.8 Concluding Remarks

Finally, it is good to point out that art therapy can be useful not only for young patients but also for their parents because, through creativity, they can achieve a living and dynamic interaction with the child, overcoming the sense of inactivity and passivity and the sense of anxiety and anger about not being able to have control and power over the well-being of one's child. Illness and hospitalization are a time of crisis not only for patients but also for the parents, because it is possible that they can often feel worthless, guilty, and hopeless.

For some of the group meetings with the children, the parents were also present, and in those moments, they exchanged opinions with each other and often the mothers spoke with the department psychologist. The psychologist and the art therapist can help the parent achieve a state of higher awareness and show them possible paths through which they can interact with the child in a more supportive manner. When parents and medical staff, especially the nurses, are adequately prepared for

art therapy, the child is able to perceive painful procedures he must undergo as being less traumatic [2].

Each time children are given the opportunity to express themselves, they will learn to be less afraid of their inner world, and they will be more likely to be able to cope with exceptional events and fearful emotions. Usually they do not aim to produce something like a preestablished result: what is important is the activity in itself. Children and adolescents use drawing and artistic expression, regardless of their specific content, to reclaim what has happened to them and to become familiar with the disease and with their fears. The experience of being able to have control over their feelings and sensations, even if on a sheet of drawing paper, is very important given that they cannot control their medical condition.

Our observations have shown that recurrent themes can easily be put in relation to the experience of the illness characteristics of young cardiac patients. In the drawings the instability and emotional fragility of children and adolescents are often clearly shown. The drawings produced mainly by teenagers seem to be in agreement with the current literature in this field, indicating the ambivalence between a desire for independence, autonomy, and wanting to leave behind their physical limitation and forms of regression and dependence on the family environment determined by the precariousness of their physical condition.

Quite often, bringing a child to the attention of an art therapy intervention is neither immediate nor simple: the hospitalized child often feels anger and fear, and his/her desire or interest is often not sufficient to understand who is in front of them and what they are proposing. The success of the implementation of an art therapy intervention cannot therefore be separated from establishing a relationship of trust previously, preferably with a single person, primarily, who should be present during the child's hospitalization and active with respect to the obvious needs of the child.

It is also important to emphasize that there can be some initial difficulties with the group dimension of art therapy, especially with children who are isolated in their rooms. These patients often have an increasing difficulty to socialize, to be interested in others; however, if they are encouraged to join the activities, it is probable that they can more effectively cope apathy, discouragement, and difficulties in general. Of course, this is subject to the physical condition of each child; through art therapy the patients can find friends who encourage the continuation of activities through individual performance in a small group.

It is important to emphasize the widespread perception of the usefulness of this type of intervention, more so in a context in which the protagonists are chronically ill children and, therefore, patients who are deprived of their daily routine. This utility is evident not only from the considerations of the parents but also – and perhaps especially – thanks to the enthusiasm of the children.

Finally, we would like to conclude this chapter with the following citation and other drawings from our laboratory (Fig. 11.5a–d):

> The creative event is a process of emotional experiences, unconscious images, conscious choices and logical structures. The creative moment is the act of expressing an emotion that comes to light. In this dynamic relationship between mind and body the space for the therapeutic relationship, rehabilitation and education is created. [19]

Fig. 11.5 Drawings from the Art Therapy Laboratory

Fig. 11.5 (continued)

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Peer-to-Peer Support for Congenital Heart Disease in Europe: ECHDO and ECHG

12

Torben Geier

12.1 Introduction

In most European countries, self-help organisations are very rampant and very different from each other. There are small groups that work only on a local or regional level and there are huge organisations which work on a national level. Some groups only work with volunteers, but there are also associations that have a lot of paid staff. The amount of money which the different groups can use for their projects also differs from some hundred Euros per year up to millions. Also the aims of the associations can be very different.

But all self-help organisations have one thing in common. They want to inform the public, politics and people who are involved in some other manner about their aims.

This chapter focuses on medical self-help groups which represent parents with children with a congenital heart disease (ConHD) and adolescents and grown-ups with ConHD as well. Like other self-help organisations which work in the medical field, peer-to-peer support is an important characteristic of these groups.

In a time in which physicians, cardiologists and even psychosocial workers in the hospitals and in medical practices only have a few minutes for treating their patients, the role of self-help groups as providers of information and support increases. The way of getting information from the group to the individual patients varies a lot. Today the groups use leaflets, booklets, books and newspapers which they publish. They have Internet pages and are linked in different social media forums. They organise scientifical symposia. But the main element of their work is still the peer-to-peer support.

Peer-to-peer support often entails that a patient with a ConHD contacts another patient with ConHD and they both speak together. The topics of these talks can have

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a lot of varieties: questions about the medical condition or therapy, about school or job problems caused through the ConHD or questions about the disability passport – just to name some issues.

The ways of communication can also differ: phone, email or Internet chat systems, meeting in person and attending group meetings. Something which characterises peer-to-peer support is that the person you contact to get assistance has gone through a similar situation and so is able to give you advice.

On a national level the information you can get from self-help groups or peer-to-peer support can be very useful for the individual patient and his/her family. Both parties speak the same language and live in the same country. This is important because both use the same social and health-care system, and so it is easy for them to compare their situations with each other and follow the advice the other one can give them.

But what happens when the peer-to-peer model is transferred on a European level? Difficulties can arise when comparing different social and health-care systems in different countries. How reliable can advice and information be, if the parties of a peer-to-peer meeting come from different European countries? With the introduction of two European umbrella organisations dealing with ConHD, the answer to this question shall be found.

The first umbrella organisation is the European Congenital Heart Disease Organisation (ECHDO), which initially was an umbrella group only for parents with children with ConHD, but within a few years of it being founded, it also included grown-ups with ConHD.

The second umbrella organisation is the European Congenital Heart Group (ECHG), a loose federation of groups that represent only adults with ConHD. ECHG has not been registered as an official European association till this date.

12.2 European Congenital Heart Disease Organisation (ECHDO)

12.2.1 History

With the improvement of cardiac diagnostic and cardiac surgery, more and more children with ConHD survived and get a chance to live. The new methods gave many children, who would have not survived otherwise because of their severe heart failure, a chace to live. A new patient group was born and also a new kind of paediatric care developed. This development started in the 1960 and it is still going on.

The great medical progress does not prevent that the diagnosis of ConHD was and still is a big shock for all parents. In the daily routine of a university hospital ward for children with ConHD and for paediatric cardiologists the contact with these patients may be standard procedure, but in public media and the public everyday life, children with ConHD live only on the fringes. Therefore, parents-to-be could have heard from ConHD in general, but they have no real understanding of this issue. They do not know the varieties of the different types of ConHD and have no knowledge about the medical options and abilities they have.

If parents-to-be hear the diagnosis ConHD for their child, they enter into complete unknown territory. The lack of information and experience can generate a lot of fears. Of course the paediatric cardiologists and cardiac surgeons can explain to the parents the concrete heart failure of their child and the next steps they have to do, but these professionals may have some difficulties to understand how the parents feel and often they are not able to dispel these fears.

Therefore, parents in Western Europe started to find national self-help associations in the mid-1970s. The first goal of these associations was to provide information about the different heart failures and the medical procedures that were available at that moment and to exchange experiences. For the first time parents with children with ConHD began to use peer-to-peer support for their work. Wherever possible parents met in small groups and spoke about their experiences, fears and hopes.

Such meetings normally took place at the hospital or nearby. However, this meant that only parents who live nearby the hospital or who have a child in the hospital could take part in these meetings. To include more parents the associations started to work with contact persons for specific regions. The contact persons were parents who volunteered to speak to other parents and share information with them. The contact details of these parents were published in the hospitals and in the newspapers of the associations, so that parents in need of advice could call the contact persons and if necessary they also met personally. Today many parents who look for information use the modern communication platforms such us emails, websites with discussion forums or social media groups. But the old ways of the direct support are still in use.

After the national self-help associations have established an exchange of information and advice, they started to work on a political basis. They tried to improve the medical care for children with ConHD and fought for more acceptance of this disease. As they saw that things in the hospitals changed and the medical care for their children really improved, the associations started to widen their political activities to other fields, like, for example, a better integration of children with ConHD into the school system or for better chances on the job market.

The paediatric cardiologists, were important allies when it comes to political demands and the provision of medical information to back up these demands. They also informed these patient groups about the pertinent medical developments and new therapies. For these reasons, the collaboration between the parent associations and the paediatric cardiologists became tighter and tighter. Representatives of the parent associations visited advanced training courses for paediatric cardiologists and cardiac surgeons to get the newest information and to stay in contact with the doctors.

More and more representatives started to visit even the events of the European societies for paediatric cardiologists and cardiac surgeons. In this way the representatives of different national self-help associations got in touch with each other. The result of this progress was that the first exchange of information between the different countries started and the first common projects were discussed.

During the meeting of the Association of European Paediatric Cardiologists (AEPC) 2006 in Belfast, the delegations from Spain, Sweden, Norway and Germany made the proposal to establish a European umbrella group for the first time named

European Congenital Heart Disease Organisation for parents of children with ConHD. All the other national self-help associations, which were present in Belfast, confirmed this proposal and also confirmed the representatives of the four countries to become part of the board.

The first ECHDO meeting was organised by the German Bundesverband Herzkranke Kinder e.V. in Berlin in January 2007. ECHDO was not an official association at that time and it was supported by the German Kompetenznetz Angeborene Herzfehler Association. The leading group presented a draft for a European project. The goal of this project was to establish a European Internet platform, on which research results about ConHD from all over Europe could be published, after editors read the abstracts and made sure they are understandable for parents.

During this first meeting, there was also a discussion about the form of the new European umbrella group. It was discussed whether the association should only be for parents or if it had to include also the associations of the Grown-Up Congenital Heart Disease (GUCH). The discussion did not come to an end in Berlin and so it became the main topic during the second meeting of ECHDO in Bruges, Belgium, in February 2007. At the end of the three days of discussion, the representatives came to the conclusion that ECHDO shall be an umbrella for all patients with ConHD and their parents.

In the years 2008 and 2009, the ECHDO meetings were held in Berlin and Barcelona. During these meetings, which both lasted three days, the delegates discussed different topics. The main concerns were the creation of statutes and by-laws for the new European association, with which the majority of represented associations could live and the developments in the EU-funded Internet platform called Corience, described further on in this chapter.

Finally ECHDO became an official European association during the first official general meeting organised by the Children's Heart Federation in 2010 in London, with the election of the first board and the last changes on the legal documents, giving rise to the first European association for patients with ConHD and their families.

12.2.2 Goals and Benefits

During the process of developing a European umbrella group, the members of ECHDO have chosen the following mission:

> ECHDO works for ConHD patients of all ages, gaining knowledge from parents of children with ConHD and from adult patients, to achieve common goals. As increasing numbers of heart children survive into adulthood, ECHDO members include both parent support groups and support organisations with a focus on GUCHs.

In its statutes ECHDO expresses the following goals:

- Improvement of the treatment and quality of life of patients with ConHD by information on appropriate medical, surgical and psychosocial therapies

- Integration of paediatric and adult cardiac treatment with the goal of installing a smooth transition for adolescents from paediatric to adult
- Promoting of scientific and medical research in all fields of ConHD
- Representing the interests of ConHD patients in national and European health policy
- Raising awareness of ConHD and instilling in the public an understanding of the concerns of people with ConHD and improving the treatments of children and adults

If you read such goals written on paper, they sound very realistic and easy to achieve. But if you try to realise them, there are suddenly a lot of problems to solve – especially if there are 36 associations from across Europe involved. Dealing with different associations from different countries is one of the greatest issues of every European umbrella group.

When it comes to the countries of Western Europe, regional or national self-help associations for parents of children with ConHD existed since the mid-1970s. These groups had a lot of members who were very experienced in raising money and public awareness for their organisations and they had some influence on the national health policy. On the other hand, there were a lot of new groups – mainly in Eastern Europe – with only some members, no or less financial support and no public awareness or political influence. Even today, in some countries there are still no working self-help groups for parents of children with ConHD.

Also the standard of treatment and quality of life for children and adults with ConHD differ from country to country. In some countries, the standard of treatment is very high and professional; in other countries, it has not reached such a professional level yet.

How can such different associations work together? The smaller associations need advice and support to strengthen their internal structure, to raise money and awareness. They have no interest in European policy at the moment – there are other problems and issues to solve first. These smaller groups would be glad to achieve what other countries already have. The bigger Western European associations are more interested in gaining influence on the European health policy and to support European research initiatives.

ECHDO has to serve both needs. It must provide information, advice and support for the associations which ask for it. At the same time ECHDO has to spend money and energy to support European research initiatives and to gain influence on decisions about health-care policy on the European level.

For the moment it seems that ECHDO is able to hold the balance between these different requirements. During the annual meeting all groups have the possibility to exchange information and the ECHDO board supports this exchange by offering lectures about different topics. Furthermore, the national self-help groups have time on such a meeting to introduce their projects and their information material to all the other groups. The result of such an exchange is that the other groups can get help for their projects or that projects in which there is the participation of various countries can commence.

On the European level ECHDO has created the website www.corience.org. This website contains information about living with ConHD and especially the newest developments in the medical field. Articles about new medical, surgical and psychosocial research results are published on this site. All articles are written for ordinary people, so that everyone can understand them. The site is available in English, Spanish and German. At the moment ECHDO participates in the European LENA project (LENA = Labeling of Enalapril from Neonates up to Adolescents), which started in November 2013.

12.2.3 Results

Despite of all the varieties of member organisations and different goals of these organisations, ECHDO is able to build up an exchange of information between the members. The best possibility for such an information exchange is the annual meeting, where the members meet in person and can introduce their projects and ideas to many other member associations.

But there is also the possibility for the members to publish information on the ECHDO Facebook page or the new ECHDO website www.echdo.eu. All these ways of cooperation are very helpful for the members, but they are not a model of peer-to-peer support, since the differences between the members are too huge.

The greater and stronger groups can give advice to the smaller ones and help them to improve their internal structure and to avoid mistakes they did during their development. With a lot of goodwill, you can see this as a part of peer-to-peer support, because all organisations have a comparable structure and way of working and so can use the advice for their own situation.

Even though there are these important differences, it is possible for the various associations to work together and to plan common projects and events. Thereby mostly organisations which are in the same situation collaborate together, because they have a lot in common and can easily find a common basis for such projects. Here the idea of peer-to-peer support is developed at most.

12.3 European Congenital Heart Disease Groups (ECHG)

12.3.1 History

The children with severe congenital heart disease who survived the first operations grew older and they reached adolescence and adulthood. Suddenly a new group of patients was born: grown-ups with congenital heart diseases (GUCHs). Even for most of the paediatric cardiologists and heart surgeons, this was surprise, also the professionals were not prepared for this situation.

But also the parent organisations did not see that there was a new group of patients with congenital heart disease. They still were focused on a better medical care for the newborns and little children. Activities for adolescences or adults were not very common, and so the GUCH patients in the different Western

European countries started to organise their own peer group activities at the end of the 1980s.

Several parent associations from different countries got an invitation to the first European holiday camp for GUCH patients in Bilthoven, Netherlands, from the 23rd to the 31st of May 1994.

The idea for such a camp originated in 1993 as two professional psychosocial workers in GUCH matters from the Netherlands (Irene van der Klis) and Scotland (Lorraine Simpson) met in a conference on GUCH matters in Vienna. During the conference in Vienna, they found out that in both countries, it was common to organise holiday weeks for GUCH patients who were not able to go on holiday on their own. So they thought about organising a holiday week for Dutch and Scottish GUCH patients in the next year. During the process of developing the idea, two GUCHs from the Netherlands extended the idea to a European holiday camp.

The first camp was visited by delegates from eight countries (Belgium, Denmark, England, Finland, Germany, Scotland, Sweden and the Netherlands). There were no lectures, no guest speakers and no workshops at all. The delegates gave a short presentation about their country in general and how they were organised as a peer group. Spare time activities, recreation and an informative exchange between the delegates were the main topics of this first European GUCH "Conference".

But at the end of the week it was clear for all delegates, that such a wonderful meeting needed a follow-up. Therefore, the delegates from Scotland invited all attendee to a further meeting in Scotland in 2 years' time.

The second conference for GUCH patients was "The European Young Hearts Conference" in Edinburgh, Scotland, from the 28th of July to the 3rd of August 1996. To finance the conference, the Scots got funds from the European "Helios II programme", which supported international projects to improve the quality of life for disabled people. In order to receive funding from this programme, there were several obligations to fulfil. One of this was that the funded project had to have an educational value. Therefore, the Scots added guest speakers, lectures and workshops to the programme and so the holiday camp which took place in 1994 was transformed into a real conference.

In 1996 12 countries took part in the conference. All countries from the former conference were present in addition to Australia, Austria, France and Switzerland.

Also in 1996 the Scottish GUCH patients founded their own self-help association "BraveHearts" and they obtained their independence from the Scottish parent organisation. This was a motivation for many other GUCH delegates who were present on the conference to go into the same direction when they returned back home.

"The Young Hearts Conference" was held in Rovaniemi, Finland, from the 5th to the 12th of July 1998, and for the first time not only the English delegates were sent from their own self-help association, but also the delegates from Austria, Germany and Scotland, now represented their own independent associations and were no longer parts of the parent associations.

The "Millennium Conference" in Oxford, England, from 3rd to 7th of July 2000 set a new record when it comes to associations and participants. There were more than 100 delegates from 16 countries. For the first time there was a lecture called "What is next for Europe" from Lorraine Simpson in which she gave the delegates

the impulse to think about getting more organised between the conferences and to develop a European vision for GUCH patients.

The lecture of Lorraine Simpson led to the "Heading for the future Conference", which was held again in Bilthoven, Netherlands, from 1st to 7th of June 2002. One of the main topics of the conference was "Advantages and benefits of having a European GUCH association". The organisers of the conference had the idea of forming a taskforce during the week that would start working to fund a European umbrella organisation for GUCH patients. But the organisers had not communicated their wish clearly enough to the different participating associations and so all delegates were surprised about the idea to form a taskforce and to start founding such a European umbrella group at once. Therefore, no decision could be made, because most of the delegates wanted to discuss the topic more deeply at home and had to get advice from the respective boards.

Two years later on the "Open Hearts Conference" in Morschach, Switzerland, from 10th to 16th of July 2004, all delegates were prepared for a discussion about forming a European umbrella group for GUCH patients. A European Young Hearts Task Force (EYHTF) was elected. The aim of the EYHTF was to write statutes for a European umbrella group and to find out how we can find it officially.

Again 2 years later on the "Open Hearts Conference II" in Skibotn, Norway, from 2nd to 8th of July 2006, the EYHTF presented their results of 2 years' work to the delegates. But there was not much to present, because after a good start, the taskforce had only had four meetings during the 2 years and most of the time discussed about their internal structure. Therefore, it was agreed that a new taskforce should be elected and that it should be given clearer and more practical tasks. The tasks of the new group were:

- To ensure the future of the conferences
- To enable communication between the conferences
- To contact and monitor progress of the European umbrella group for parents that started to form

The taskforce was now named European Congenital Heart disease Groups (ECHG).

After Norway the conferences returned to Finland where was held the second "Young Hearts Conference" in Tampere from 13th to 19th of July 2008. The ECHG committee fulfilled its tasks by publishing guidelines with the title "How to organize a conference", a website was created and members of the committee visited the first meetings of the parent organisation ECHDO in Berlin and Bruges. Furthermore, the committee wrote statutes for its election, internal structure and internal cooperation.

This way of working was a real success so that a new ECHG committee was elected and had the following tasks, for the next 2 years:

- Maintain and develop the website
- Maintain and strengthen the relationship with ECHDO
- Support for the conference organisers
- Develop a guide to GUCH organisations

- Search ways of funding the conferences and the ECHG committee
- Develop statutes for the ECHG umbrella group

In 2010 the conference was held from 19th to 23rd of July in Berlin, Germany, under the title "EuroHearts-Conference". The ECHG committee completed most of its tasks. It developed a new guide "How to found and run a GUCH organisation", produced a leaflet about the ECHG for possible sponsors and created a good relationship with ECHDO. The website of the ECHG still existed, but only one new article was published between the conferences. The committee worked on their internal statues and send out a questionnaire to all participating countries of 2008 to get ideas for the statutes of the European umbrella group that should be founded in the future. But only two European countries answered the questionnaire and so the committee was not able to write the statutes for the umbrella group.

But nonetheless the way of working was still good and so a new ECHG committee was elected and got the following tasks:

- Maintaining and updating the website
- Fundraising
- More help for countries organising the conferences
- Creating a history of the conferences
- Maintaining contact to ECHDO
- Building up a new structure and a better communication between the conferences

From 30th of July to 3rd of August 2012, the "EuroHearts-Conference – Generations of Hearts" was held in Copenhagen, Denmark. The ECHG committee could present the "History of the GUCH conferences 1994–2010". The communications has been approved by a new Facebook group www.facebook.com/ECHGroup and the installation of a newsletter. Contact persons from the different countries sent articles about the activities in their associations to the committee which then wrote the newsletter and sent it out to all participating associations. But the website of the ECHG was closed, because more and more communication and traffic ran through Facebook.

There was so much work to do for the committee that for the first time a personal meeting of the committee was organised in London in December 2011.

In Denmark a new ECHG committee was elected with the tasks to maintain the communication between the conferences and to write a cost-benefit analysis about founding an own European umbrella group for GUCH patients.

The next conference will be held in Baar, Switzerland, from 20th to 25th of July 2014. So the history of the GUCH conferences and the ECHG committee continues.

12.3.2 Goals and Benefits

As can be seen in the history of the ECHG, the meetings started as an experiment and in the beginning the only goal was to get in touch with other adults with ConHD. However with time, the GUCH patients got more and more self-confident. Building

up new friendships was no longer the only goal of the conferences. The patients started to exchange information about their countries, the health systems and the culture of self-help associations. They also spoke about medical, social and psychological issues concerned with ConHD and so they got the courage to get even further and try to found their own European umbrella group.

The ECHG still is not an official European umbrella group, however a clear vision has been developed. The objective it to create a world in which children, adolescents and adults with a ConHD have the same chances for attending schools, studying and getting a job like everyone else.

The people working for the board of ECHG want to achieve this vision through campaigning. They want to show the public what people with a ConHD can achieve and that they are as valuable for society as healthy people. But to achieve this on a European level, they have to get stronger and better organised. Therefore, they founded the ECHG which makes sure that the conferences go on and more and more countries can become the host of it. The ECHG supports smaller groups in building up their internal structure to get stronger and to gain more visibility in their home country.

The benefits of the ECHG so far are that they make sure that the conferences will go on and that there will be a constant exchange between GUCH patients from all over Europe on a regular basis. Furthermore, friendships that started on the conferences hold until today, because of the new media. Social media give people a very good opportunity to stay in touch with each other in the months without a conference. It is a good platform to exchange ideas and information overcoming long distances, different time zones and other obstacles. In this way the GUCH community will become more united.

Even people that have never been to one of the conferences and maybe never will have the opportunity to travel to such a conference can get in touch with the delegates and the members of the ECHG working group. The exchange between the patients and the national associations grows and many issues can be discussed before and after a conference so that long-term goals can be defined and reached.

12.3.3 Results

Like ECHDO the ECHG has too few personal meetings in order to enable adequate peer-to-peer support. The members of the ECHG meet on a biannual basis and in each meeting a lot of delegates have changed. The advantage is that a lot of new knowledge and a lot of different experience enrich the community. On the other hand the delegates have to build up a new trustful relationship before they can exchange information and experience.

The ECHG conferences – like the ECHDO general meetings – can only give an impulse for the participating organisations for common projects. The main work has to be done between the conferences and meetings. The ECHG committee – like the ECHDO board – has the rule to coordinate common projects, fulfil the tasks the community has given to it and ensure the flow of communication.

The main work and communication in both networks runs over the Internet. Websites, emails, social media and the opportunity of communication software like Skype give every association and even every individual the possibility to exchange information, ask for advice and receive it. This way of peer-to-peer support is not the classical one everyone knows – it is a new kind of assistance.

This new way of peer-to-peer support may not be as efficient as a personal meeting of the peers, but also on the national and regional level sometimes this is not possible, where telephone calls, emails and social media are also instruments of communication and peer-to-peer support.

Therefore, the way of the ECHG and ECHDO is an appropriate instrument to exchange information and advice between the different associations and countries. Peers from all over Europe can get information from every other European country and use it for their own situation.

Conclusion

Both groups started from *very different starting points.* The parent associations – especially in Western Europe – had a long history and a lot of experience in working as a self-help association and with peer-to-peer support as they became an official European umbrella organisation in 2006. The GUCH patients were not organised as they met on their first conference in 1994. For them founding and running a self-help organisation and providing peer-to-peer support were still distant objectives. They knew the parents did such things for their members, but for the GUCH patients of the first generation, it was unimaginable to have such services, too. First of all they had to build up self-help associations and peer-to-peer support in their own countries.

Despite of these different starting points, both groups have independently developed a similar way of working. Both use the Internet for the main communication and share a lot of information through emails and social media bulletin boards.

These instruments help to overcome the distance between the different associations and gave them the feeling of being one community. The different healthcare systems in each country make it necessary that the information from other countries cannot be used one by one but have to be adapted to their own circumstances. However this does not prevent the provision of peer-to-peer support from happening completely.

Even the biggest obstacle – the language barrier – does not stop the exchange of information. Spanish comics are translated into German, Norwegian brochures are available in Russian and there are many other initiatives which are being replicated.

Peer-to-peer support is also a success on a European level, and at the moment there is an attempt from both ECHDO and ECHG try to find out how they could combine their knowledge and experience and how they could cooperate more efficiently. The next step to improve the quality of life of all patients with a congenital heart disease has begun.

Suggested Reading

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The Role of Associations in Congenital Heart Disease: Peer Counseling and Advocacy

13

Giovanna Campioni, Edward Callus, and Emilia Quadri

13.1 Associations in Italy

In recent years, the number of voluntary, nonprofit associations formed by patients, created around the issues of a given disease, has increased. The formation of these associations normally arises from the need to meet, more broadly, some services provided by the national health system and to carry out initiatives for the protection and health and social care.

In many cases, the complexity of the situation has led to a progressive organization of individual associations getting together and forming federations to give more strength to their message – both nationally and internationally – without losing their individual characteristics. In this way, requests for good quality care are becoming stronger and equally available nationwide, as well the dissemination of information about diseases and possibly their prevention or early detection. This also creates favorable conditions for investment in scientific research to improve the knowledge about diseases and to protect the patients and their families.

In fact, the demand by citizens and patients to play an active role in the choices on health, both in the doctor-patient relationship – that is, increasingly moving away from a paternalistic vision of the role of the physician and it is becoming a relationship in which choices are shared – and also in health policy and research, has been growing, for several years [1].

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These needs were expressed initially by the movements for the protection of citizens "rights and patients" associations. The first to be created and grow in the last century have been women's associations involved in the fight against breast cancer in the United States and the movement of the fight against AIDS, a disease with a strong social impact as well as one on health [1].

To these movements "historic" associations over time associations that deal with various diseases all over the world were added. Most often, the objective of these associations is to help their members find solutions to everyday life problems of the disease in pragmatic ways, such as providing technical and psychological support to the patients and their families (and also to society). Nowadays these associations are broadening the range of their influence, by disseminating information on the illnesses (education), by participating in and orienting clinical research, and by influencing health and social policy on the basis of the needs of the patients also through lobbying activities.

It should also be pointed out that the health services at the international level are going through a critical period in terms of financial sustainability, which has been accompanied by the spread of patients' associations and citizens that make up for welfare services which would be difficult to obtain in the healthcare system and promote awareness-raising initiatives so as the rights and needs of those they represent are respected.

The role of patients' associations is therefore essential in order to promote and improve the dialogue between doctor and patient, as well as to encourage communication and discussion between the individual patients, in order to clarify the needs and expectations for a better understanding of the developments in the medical and scientific fields, with the ultimate aim of improving the quality of life of these patients and avoid the so-called social selection.

Patients' associations are a key reference point for those who suffer from a specific disease. They give support and information, represent the point of view of patients, and organize awareness campaigns on issues that affect their lives and those of their families. Some of them are also engaged in promoting research into the causes and potential treatments of specific diseases.

In Italy, among the emergencies of the national healthcare system, there is now the issue of adults with congenital heart disease (ACHD). These people, born with a congenital heart defect, underwent cardiac surgery as soon as they were born or during childhood, have passed the critical stage of adolescence, but are having to deal with a number of health, psychological, and social problems, related to their medical history, as also outlined by the literature [2, 3]

ACHD associations are necessary because, in part they make up for an institutional lack. For example, when it comes to the Italian national health system, not all Italian children with congenital heart disease were operated in their territory of origin, most had to travel long distances to move in specialized centers. When they returned home, even if they were followed by the medical center of reference for many years, in most cases, these children did not find a proper medical support in their city or town of origin in. They grew up, together with their parents, mostly day by day, not knowing how to deal with even the most trivial problems. When they

became adults they had no effective recognition of their pathology, and they were not helped to find a job. Compared to their healthy peers, they had a much more difficult life, and they had to solve problems such as studying, work, pregnancy, etc., virtually alone, as also outlined by the qualitative study on life experiences carried out in our country [4].

Being part of an association can help these people, not only to live better because there is the possibility to keep updated to the constant flow of information, the possibility to contact psychologists and medical doctors but also to overcome isolation and in being part of a group that works with common goals, which can serve to make citizens more aware of their rights, not to mention the right to happiness. It has been highlighted in the pertinent literature that this population is interested in receiving long-distance support and also peer-to-peer support [5], and associations are excellent at giving such kind of support.

13.2 AICCA ONLUS: The Italian Association of Adults with Congenital Heart Disease (Associazione Italiana dei Cardiopatici Congeniti Adulti)

The Italian GUCH Association (AICCA, Associazione Italiana dei Cardiopatici Congeniti Adulti ONLUS) was founded in 2009 from the desire of people with congenital heart disease to create a support network which acts on different fronts, and it includes people with congenital heart disease, their families, physicians, surgeons, psychologists, and peer counselors, specialized in caring for people with congenital heart disease and their families. In Italy, there are over 100,000 people with congenital heart disease but this pathology is not known at all on a public opinion level. AICCA would like to assist this population and help them to deal with their problems. In fact, the growing number of those who are born with a congenital heart disease and who reach an adult age is creating a situation of medical and psychological emergency.

AICCA is coordinated by young adults born with congenital heart disease: their role is to be the leaders and actors in the projects aimed to help other people with this condition.

In fact, for the first time in Italy, these young adults have taken on this challenge collaborating directly with the health professionals in order to find efficient solutions together and to propose new challenges in scientific research.

The objective is to prevent the manifestation of medical and psychological problems through the thorough knowledge of the main healthcare techniques inherent to the patients' own condition and pathology.

For AICCA it is important to get to know the problems which are present during adulthood not only to enact help strategies but also to support parents who have a child with congenital heart disease in the different life phases of psychological and physical development of their child.

AICCA proposes to develop whichever activity is necessary or beneficial in order to improve the quality of life of adults born with congenital heart disease and

their families, creating moreover a national support network managed by young adults with congenital heart disease.

This network should allow to share similar experiences and advice when it comes to emotional, psychological, and practical issues on all things related to daily life (e.g., insurance, occupation, social services, contraception, sport); at the same time, another one of AICCA's objectives is to be an important reference point when it comes to the medical point of view filling in the gap there currently is in the Italian healthcare system which necessitates to be further capable of being able to care for people with congenital heart disease, also during their adult life.

Currently, AICCA is present on the national territory with regional branches in Lombardy, Sardinia, Sicily, and Puglia. AICCA is in touch daily with its members in all Italy though the website www.aicca.eu and the main social networks (Facebook and Twitter).

To date AICCA is dedicated to the realization of several projects including:

- The creation of a website which facilitates the communication of information, the exchange of the different points of view, and the story of experiences and encourages contact with specialists in health-related, psychological, and legal issues among patients residing in different cities of Italy.
- It is committed to increasing the regional offices in order to make the patient feel as close as possible to the mother association.
- Provides each member a health passport stored on a USB, in order to make it safer and less cumbersome, moving to other cities, nations, or states.
- Organizes an art therapy laboratory within the department of pediatric cardiac surgery at the hospital in the IRCCS Policlinico University hospital in Milan, which is described extensively in another chapter in this manuscript.
- Gives support, through its volunteers, patients and relatives, psychologists, peer counselors, to children, parents, and relatives during the pre- and postoperative period.
- It focuses on the individual, fostering the relationship and collaboration between physicians, nurses, and psychologists, for the protection and the humanity of the hospitalized people.
- Concretely supports families who are in a condition of hardship and are encountering difficulties due to the remoteness of their city or country and who are in a state of economic need, offering them hospitality, a landmark, a cozy place, and moral support to make this experience less difficult.
- The creation of a transition clinic, in order to facilitate the transition of adolescent patients with congenital heart disease to adulthood. The project involves psychologists, doctors, and nurses.
- Collaborates with several magazines in order to provide a tool for continuous adjournment in the pertinent areas.
- It is dedicated to the dissemination and increased awareness in congenital heart disease by organizing and participating in events, both medical and otherwise, with the aim of becoming a reference point not only for those who have already had problems but also and especially for those who unfortunately will have similar problems, giving them the knowledge that a good and rewarding life is possible.

AICCA wants to help turn around a difficult experience which marks the lives of so many children and their families in a positive resource, assisting and helping with each other to overcome the problems in adolescence and reaching the adult age aware of their limitations but also of their amazing and unique individual resources.

At the heart of AICCA are patients and relatives who have decided to share their life experiences, encouraging communication with those who, like them, are experiencing frequent hospitalizations.

Knowing the testimonies and experiences of those who work in the association is important because it highlights the activities and possible actions to trigger positive changes and not live the disease passively. Getting to people who have gone through or are going through the same difficulties makes you feel less alone. Meeting people who have overcome the same problems offers hope and optimism.

These are the principles that move me every day. I was born with a heart defect, which was diagnosed 15 days after my birth. I was born with a single ventricle, and this resulted in ongoing and lengthy hospitalizations, that led me to be operated four times in the heart. This does not stop me from living a life which is similar to others and to be able to have an optimal quality of life, respecting my limits and my needs.

I started working for AICCA since 2009, the year of its foundation, holding the position of national coordinator. Besides the typical bureaucratic work of the association, I realized that there was a need for me to work directly in the field, close to the patients born with cardiac malformation and their families.

This awareness has led us to understand with psychologists the need to be prepared to be able to operate properly in this field, and for this reason, I followed a 3-year training in counseling. Even though there are examples of patient advocacy and peer-to-peer specialized support, the figure of peer counselor is not so widespread in Italy.

13.3 Counseling and Peer Counseling

The counselor is a professional who, after having obtained a diploma to following a course of studies of at least 3 years, is able to facilitate the resolution of existential hardships. They are "professional listeners" who do not give advice, but competence and understanding. The counselor is a facilitator in the helping relationship, representing a reference for those who, finding themselves in a difficult moment, feel the need to be accepted and heard by someone competent and willing to take care of them. They are an expert in:

- Communication
- Hospitality
- Problem solving
- Decision-making [6]

Counseling is based on the original intuition of Rogers, according to which, if a person needs help, the best way to come to his/her aid is not to tell her/him what to

do (paying attention to the intellectual content of this advice, i.e., truly "sensible" compared to the situation) but rather to help him/her understand his/her situation and handle the problem alone and assuming full responsibility for any of his/her choices.

On the other hand, what characterizes peer counseling is a similar life experience of the consultant. This element is a priority because it enables "mirroring" in a spontaneous way, that is, an initial form of identification that is the major stimulus to undertake the counseling process. In this sense, the more similar the disability is – for example, the severity, the type, etc., the more intense the relationship and the level of reciprocity will be. In addition, it will be easier for the person being helped to be more open and to let his/her defenses down.

Peer counseling was initiated in the United States in the mid-1960s, when a group of UC Berkeley students with disabilities, the pioneers of the movement "Independent Living," agree to meet regularly and to devote time each other to discuss and exchange views on practical and social issues that they were dealing with every day. In these groups, they offered each other mutual emotional support, and through an internal process and one regarding the growth of the group, they experienced the strength to contribute to the change of the condition of life of the disabled people.

Currently in the United States, peer counseling is regarded as the most well-known forms of support and therapy. Gradually, in the 1980s, the method of peer counseling is brought to Europe where it finds fertile ground to grow and evolve. In Europe, its value is recognized within the disability movement but not by official bodies and the government [1].

The American model has been taken up in Europe where there is a network of experiences in the field of disability. This refers primarily to the initiatives related to the independent living movement, substantially DPI (Disabled Peoples' International, http://www.dpi.org/) and ENIL (European Network for Independent Living, http://www.enil.eu/), which were both initiated in Germany, where there is the most significant network of peer counseling services [1].

In Sweden, Ireland, and Germany, peer counseling is practiced in the Centers for Independent Living, and in Finland it is practiced informally in associations and is based mainly in giving "practical advice" (peer support). The Netherlands is the only country where the practice of peer counseling is done in private, through a special foundation, as freelancers. In France it is called "emulation adviser" emphasizing the importance of "role model" that is the living example that the counselor represents [1].

In Italy, peer counseling is absolutely not taken into account by the institutions, while its value is highly recognized by the movement of people with disabilities, because the latter have found that through the application and use of this method, it is possible to carry out a personal project of independent living.

In the year 2000, a second-level training course funded by the province of Catanzaro was organized by DPI Italy. Since 2001 DPI Italy has a peer counseling service at the University of Calabria.

In 2005, DPI Italy published the book *Peer Counselling: From Victims to Protagonists of the History of Life* [1]. The book, published in Italian and English, is already in its second printing.

At an institutional level, with the Decree of the President of the Council on April 9, 2001, the "Uniformity of treatment on the right to higher education, in accordance with Article 4 of the Law of 2nd of December 1991 n. 390," registered at the Court of Auditors on 6.6.2001 (reg. 7 fg. 173), Art. 14, with regard to interventions for students with disabilities (paragraph 10), it is textually stated: "… The interventions of mentoring can also be entrusted to "peer advisors", ie people with disabilities who have already faced and solved similar problems to those of whom turn to ask for support."

Peer counseling is a special form of counseling, initiated from the experience of associations and persons with disabilities, to assist the process of individual empowerment and, consequently, of social emancipation. It allows to break the "cast" that has settled around the authentic core of the person, which prevents their expression and development.

For the person with a disability, breaking these boundaries involves a complex process, including the overcoming of some basic obstacles such as the historical weight of a culture of rejection and marginalization of diversity, made up of stereotypes still present at the level of social and interpersonal communication, for which we are still paying the consequences; the individual experience determined by personal history. Only by doing this the person with disability can live in freedom, and basing himself/herself on personal tastes and choices, and on one's own way of being.

Depending on individual needs, peer counseling may allow the people who are followed a reversal of the management of their own lives. Similarly it can help accomplish an adjustment or even embark on a new path which affects the person's personal future.

The process that is developed, thanks to peer counseling, leads to the acquisition of self-awareness, and therefore, the use of these internal instruments – psychological, emotional, imaginative, rational abilities – and external ones – such as human aid, laws, assistive devices, technologies, etc. – fosters self-realization.

The issues faced in peer counseling and the goals that are pursued may be the most diverse. It begins with the sharing of the experience of disability and covers issues such as the management of personal assistance, discrimination at the workplace, and problems of mobility. But almost always, the specific arguments serve as a stimulus to bring to light the whole personal world including personal experiences, relationships, and emotions, starting with the "here and now."

Through the method of peer counseling, people are helped in increasing their ability to move from a situation of learned passivity, which causes feelings of helplessness in the face of alienating or frustrating experiences to the learning of hope, which is derived from the feeling of increased control over events, through participation and commitment in the context of their own lives.

13.4 Congenital Heart Disease Patients: Why Is Peer Counseling Important?

As specified earlier in this chapter, people born with a congenital heart defect, in the course of life, are confronted with a series of health, psychological, and social problems, related to their medical history.

These people have to solve very difficult problems related to their studies, work, pregnancy, etc., practically alone. Many of them lack the awareness of the real health state [7] while ignoring their potential resources. In most of the cases, there is an emphasis of the things they cannot do, at the expense of expressing their full personal potential.

This suggests the need for a specific approach; the peer counselor is the figure that through collaboration with psychologists can contribute to addressing the disease and get answers to their questions.

Over the years, this innovative approach has given significant results, both for the patients themselves and for their families. Thanks to this type of intervention, family members manage to be more optimistic with regard to their loved ones. They can verify that other kids, with heart defects similar to those of children, were able to reach important achievements in life, to pursue their aspirations, and to realize their dreams. The results of greatest satisfaction have been seen specifically in people with congenital heart disease.

Many patients are able to see that, by accepting their limitations and difficulties, nothing prevents them from having a fulfilling life. They learned to live, not just survive, and they understood that there are many more things that you can do that what cannot be done. As a national coordinator of an association for adults with congenital heart disease, I have had the occasions to provide peer-to-peer support to many patients. I have accompanied them on their journey, and together, for each one, we have found the best path to follow.

13.5 A Case Study of Peer Counseling in Congenital Heart Disease

At the time I met him, S. was 40 years old and he has always considered his life as being different, but mostly he was treated as someone who was ill.

S. lives in a small town in Sicily and is the eldest of three children. His parents were always overprotective toward him; in fact they never supported him and encouraged him to study, because they believed he was not capable to do so due to his serious health conditions. Eventually the fears of his parents also became his fears, and mainly because of this, he never had a girlfriend and at his age he had never had sexual encounters.

I met S. during one of his hospitalizations, and when he got to know about the possibility of me providing peer-to-peer support, he immediately asked me if this was possible. Since he lived far away, we arranged to have telephone/Skype calls once a week for 8 months. S. had many issues to work through, but the fact of

having the same health condition, and the possibility of providing peer-to-peer support, greatly facilitated the work.

After 8 months, S. is more aware of his medical condition; he learned to understand what he can and cannot do, greatly improving his self-esteem. He also found a girlfriend, and finally, at 40 years, he had his first sexual intercourse.

13.6 Advocacy

Disability in itself gives rise to a situation of disadvantage. The disadvantage is caused not only by the limits imposed by the disease, which consequently create the situation of disability, but also by the social, cultural, and urban environment in which the person lives. Also the environment causes situations of disability, limiting the sphere of experience and compromising the natural path of life of the person with disabilities.

The path of the life of a child for whom the parents do projects – educating him/her to be autonomous, accompanying the growth process, and facilitating the meeting with peers through playing, in places such as school education – undergoes drastic changes if the child is born with a disability. As a consequence of this, the child is to be considered a person to protect, take care of, for whom they will be responsible all their life; often the concern of the future takes over. Even the usual places of everyday activities are transformed into places in which one needs to pay attention; there is no project for him or her to create a future where he or she is autonomous, because the goal becomes to achieve healing.

The lack of socializing experiences, overprotection, and always being around to people who think, speak, and decide for you are factors that cause the disruption of normal growth path.

A true work opportunity is based on the concept that the disability becomes a resource. Careers that people with disabilities and their families live every day, also with the presence of frequent clichés and the daily efforts of facing the convoluted bureaucracy to obtain a minimum of dignified existence. And it is at this stage that the encounter with other people with disabilities, parents of other disabled people, organizations, and persons with disabilities occurs.

The first approaches have as an objective providing information and offering help, in some cases, the supply of services, including rehabilitation. The meeting and discussion with those who have already lived through marginalization situations, the consequences of a past which is too muffled and protected, highlight the need that in order to get out of this situation, there is a necessity of re-elaborating one's life story and to learn to rediscover, accept, and love oneself, a process essential to face life with determination.

Gradually the concept of "right," in terms of legitimacy, is obtained, initially with great confusion, passing through resignation because initially there is the impression that nothing is due to the fact that there are fundamental rights. Only at a second moment, and thanks to an effective information, support, and a process of

empowerment and advocacy, the awareness of the "right and duty" and, therefore, active and aware citizenship is obtained.

In Italy, the term "advocacy" is translated as to "the protection of the rights of vulnerable groups of the population." In this sense one speaks explicitly of "voluntary human rights" [8].

Advocacy is very present in European and international associations and it comes in different forms, depending on the area and its needs, cultural influences, and transferred experiences.

In the world of disability, advocacy work is done within services which are almost always organized by voluntary or nonprofit associations, and in many cases, the same advocate is a person with disabilities, and he or she is a professional working in the social sector and does not work to win cases, but works toward achieving rights.

The advocate is someone who acts as an advisor inside information services, and their job does not only entail the mere dissemination of useful information but also helping the person with a disability to acquire all of the necessary tools to make sure that their rights are guaranteed, providing support if there are any cases where the disabled person's rights are violated.

The role of the advocate is more commonly known as support, or peer support if conducted by a person who has similar problems to the patient being supported (e.g., if the support is aimed at people with disabilities and the person providing professional support has the same disability) and as a peer counselor (if the form of counseling is closer to a helping and supportive relationship). This role is not to be considered as a substitute, but one of accompanying and finalized at supporting the person to become more independent and free in his/her choices.

The advocacy group is organized by associations or different kinds of organizations which are going through different struggles in order to claim rights. Since they are political and social actions aimed at the common good, advocacy groups perform as active subjects of representation, stakeholders, and advocates of social causes. One can struggle to obtain a law, services that improve people's lives, instead of others who marginalize. The most important thing is the involvement of the people who are directly interested in the struggle or in the campaign to obtain rights.

For this reason in our association, AICCA, we have an interdisciplinary staff which can respond to all enquiries in a comprehensive way.

The medical staff responds to all possible questions of patients from the scientific point of view: building a more direct and immediate relationship with a team of cardiologists and cardiac surgeons who can "be there" for the patient always, not just during the monitoring visit and serious situations. Living well with heart disease also means to know one's condition thoroughly and to know exactly how to behave in every situation of life. The Scientific Committee of AICCA aims to create brochures, publications, and information initiatives addressed to the members, but not only to allow for the more in-depth knowledge of congenital heart disease.

Even psychologists play a crucial role: by asking our members what they expect from AICCA, the first response was "psychological support," a need which is

experienced not only by those who are in hospital to deal with an operation or catheterization but also by those who are well. This is because growing up, facing the uncertainty of the future, and establishing relationships with others are difficult tasks for everyone, but especially for those who must live with a heart which is "not perfect." This is why the presence of a team of specialized psychologists is critical to an association such as AICCA; often those who live in a situation of distress do not know who to turn to and who to ask for help because they feel guilty, ashamed, or afraid to not be understood.

AICCA also has an objective to create training programs for medical doctors and the nursing staff so as to improve the assistance to congenital heart patients. This kind of training has already commenced in the psychological area, and AICCA offers the possibility of practicums in psychology and psychotherapy in the field of problems linked to congenital heart disease.

There is a lot of work to be done and it can be very tiresome; however, thanks to the efforts of all the different professionals who make up the association, things are evolving and changing: a lot has been done, but there is still a lot more which needs to be taken care of. AICCA will do its utmost and we hope to receive more support so as to be able to strive for our mission with increasing verve.

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