

Quality of life of grown-up congenital heart disease patients after congenital cardiac surgery[☆]

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Abstract

Background: Due to better early and long-term outcome, the increasing population of grown-ups with congenital heart disease (GUCH) brings up unexpected quality of life (QoL) issues. The cardiac lesion by itself is not always the major problem for these patients, since issues pertaining to QoL and psychosocial aspects often predominate. This study analyses the QoL of GUCH patients after cardiac surgery and the possible impact of medical and psychosocial complications. **Patients and methods:** A questionnaire package containing the SF-36 health survey (health related QoL), the HADS test (anxiety/depression aspects) and an additional disease specific questionnaire was sent to 345 patients (mean 26 ± 11 years) operated for isolated transposition of the great arteries (TGA), tetralogy of Fallot (TOF), and ventricular septal defect (VSD). The scores were compared with age- and gender-matched standard population data and in relation to the underlying congenital heart disease (CHD). **Results:** In all SF-36 and HADS health dimensions the GUCH patients showed excellent scores (116 ± 20), which are comparable to the standard population (100 ± 15), regardless of the initial CHD ($p = 0.12$). Eighty-two percent of the patients were found to be in NYHA class I and 83% patients declared that they do not consider their QoL to be limited by their malformation. Complications like reoperations ($p = 0.21$) and arrhythmias ($p = 0.10$) do not show significant impact on the QoL. The additional questionnaire revealed that 76% of adult patients have a fulltime job, 18% receive a full or partial disability pension, 21% reported problems with insurances, most of them regarding health insurances (67%), and 4.4% of adult patients declared to have renounced the idea of having children due to their cardiac malformation. **Conclusion:** QoL in GUCH patients following surgical repair of isolated TOF, TGA and VSD is excellent and comparable to standard population, this without significant difference between the diagnosis groups. However, these patients are exposed to a high rate of complications and special psychosocial problems, which are not assessed by standardized questionnaires, such as the SF-36 and HADS. These findings highlight the great importance for a multidisciplinary and specialized follow-up for an adequate management of these complex patients.

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1. Background

Prior to the advent of surgical treatment for congenital heart malformations in the 1960s, less than 20% of children born with such a lesion survived to adult life [1]. Now, 85% reach adulthood and the majority of deaths from congenital heart disease (CHD) occur beyond 20 years of age [2].

Due to the success of pediatric cardiology and cardiac surgery over the last three decades, the number of grown-up patients with congenital heart disease (GUCH) has been growing steadily, so that in the year 2000, the number of adult patients was roughly equal to those under pediatric

care. Twenty years from now, demographics forecast predict, that there will be even a noticeable larger number of GUCH patients than children with CHD [3]. Consequently, the aim of surgical therapy has changed from only a therapy to survival towards offering corrections allowing an almost normal life in terms of expectancy and quality. It is apparent that assessments of mortality and morbidity are too narrow to analyze the benefits of therapies thus additional outcomes of care are also important. The grown-up congenital heart patient is not simply a larger subject compared to the GUCH child subject. The cardiac lesion by itself is not always the major problem for these patients, since issues pertaining to QoL and psychosocial aspects often predominate. Therefore, interest in the QoL among GUCH patients is increasing in order to design, adapt and optimize the specific needs and complex medical, surgical and psychosocial follow-up of this special group of patients.

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The aim of the study was to analyze QoL, as well as specific psychosocial problems encountered in the daily life described by GUCH-patients following correction of isolated TGA, TOF or VSD.

2. Patients and methods

A questionnaire package was sent to 345 patients in French (44 patients) or in German (301 patients) according to their most familiar language. The questionnaire package contained: the SF-36 health survey, the HADS test and an additional questionnaire assessing more specific questions of GUCH patients' daily life.

3. Data collection instruments

3.1. SF-36

The SF-36 belongs to the generic measures of QoL and fulfills stringent criteria of reliability and validity [4,5]. It contains 36 items and uses a multiple item scale to assess eight emotional and physical concepts. Raw points are transformed into scores ranging from 0 to 100 for each dimension: physical functioning, role limitations caused by physical health problems, general health, bodily pain, role limitations caused by emotional problems, social functioning, energy/vitality and mental health.

3.2. Hospital anxiety and depression scale (HADS)

The HADS focuses on the two aspects of emotional disorders, which have the most clinical relevance, i.e. anxiety and depression [6]. It consists of 14 items, of which seven reflect anxiety, and seven depression, respectively. Each item is answered on a four-point (0–3) scale response category. The possible scores range from 0 to 21 for anxiety and 0–21 for depression. Each mood state can then be divided into four categories: normal, mild, moderate and severe.

3.3. Additional questionnaire

An additional disease specific questionnaire was sent to all patients focusing on demographic and social areas, medical situation and care, pregnancy, and aspects of daily life.

3.4. Patients

All patients operated at the University Hospital Berne for isolated TGA, TOF and VSD who had at least a 10-year follow-up were eligible for the study. Patients suffering from additional chromosomal aberration were supposed to create a selection bias and potentially not able to answer the questionnaire, and were therefore excluded. Patients were initially divided into three groups, according to the isolated underlying CHD: TOF, TGA and VSD. This division permitted comparison of cyanotic with acyanotic conditions, as well as evaluation of patients considered to be surgically cured, corrected or palliated, and the categorization according to the GUCH risk group classification.

3.5. Statistical analysis

The SF-36 and the HADS test were analyzed in accordance to their manuals. Scores were adjusted for sex and age in order to be comparable with standard population. The statistical analysis was performed using SPSS 15.0 software (SPSS Inc. Chicago, IL). Descriptive data are presented as means \pm standard deviation.

For comparisons of categorical items per groups χ^2 -tests were performed, and for all other data, metric or ordinal, Kruskal–Wallis tests, or, for comparison of two groups, Mann–Whitney tests were calculated. The correlations have been calculated using Pearson's product–moment correlations.

$p < 0.05$ was considered significant.

4. Results

4.1. Analysis of the collective patient group

Out of the questionnaires sent to 345 eligible patients, a total of 153 (44%) were returned and filled out correctly (TOF, $n = 43$; TGA, $n = 59$; and VSD, $n = 51$), permitting investigation of social and demographic data and assessment of QoL. The mean age of the total collective was 26 ± 11 years, 29 ± 14 years in the TOF-, 23 ± 7 years in the TGA, and 27 ± 10 years in the VSD group. Ninety-four males (61%) and 59 females (39%) were equally distributed within the three groups.

With regard to the overall QoL, all groups were found to have similar or even higher scores in all SF-36 (116 ± 20) and HADS health dimensions compared to a standard population (100 ± 15) matched for age and gender (Fig. 1, Table 1). Overall, there were few differences in the eight health

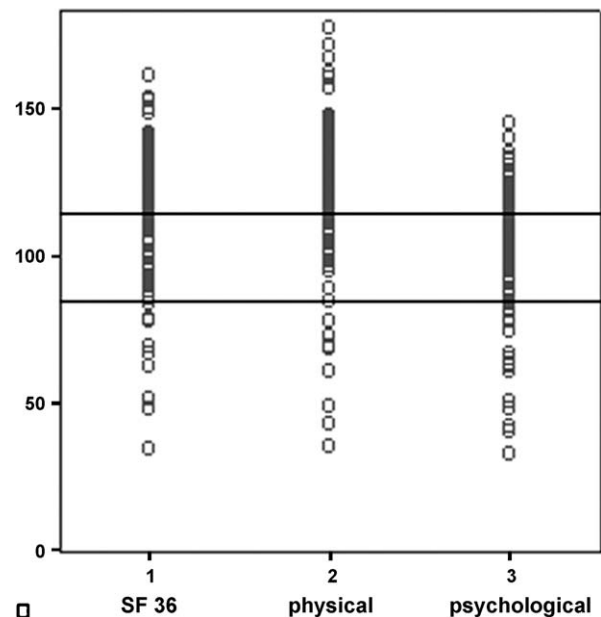


Fig. 1. Overall scores of QoL (normal range: between 85 and 115). Overall QoL for GUCH patients was excellent, even superior to the standard population in all physical and psychological aspects of the SF-36 questionnaire.

Table 1
SF-36 mean scores for each diagnosis group.

	TOF	TGA	VSD
Physical functioning	124.2 ± 33.7	117.4 ± 25.5	125.4 ± 25.1
Physical role functioning	134.8 ± 29.3	120.2 ± 35.2	125.7 ± 31.3
Bodily pain	129.3 ± 24.3	124 ± 27.1	130.2 ± 25
General health	135.9 ± 27.6	122.6 ± 33.4	134.7 ± 34.4
Vitality	114.2 ± 35	104.6 ± 29.2	108.6 ± 26.1
Social functioning	111. ± 13	104.1 ± 22.2	109 ± 15
Emotional role functioning	106 ± 25.2	98.7 ± 33.3	102.6 ± 31.1
Mental health	107 ± 22.3	99.9 ± 21.7	104.6 ± 15.3

Normal range: 85–115.

related dimensions of QoL ($p = 0.12$) (Fig. 2), and for the scores of anxiety ($p = 0.32$) and depression ($p = 0.17$) (HADS) between the three groups (Fig. 3).

Age did not affect the overall QoL ($p = 0.81$), but affects the general health ($p = 0.002$), vitality ($p = 0.001$), psychological functioning ($p = 0.005$), quality of life ($p = 0.007$) (SF-36) and the anxiety ($p = 0.008$) and depression ($p = 0.002$) scores (HADS) of the TGA group, which showed significantly worse scores with the increasing age when compared with those of VSD and TOF patients.

Analyzing the impact of reoperations, it was found, that patients requiring one or more surgical interventions (32%) (mean: 1.4 operation, range: 1–3) did not show lower QoL scores than non-reoperated patients (68%) ($p = 0.21$) (Fig. 4). Sixty-six percent of the TGA, 39% of the TOF and 10% of the VSD patients underwent reoperations, of which the TGA patients being the group with more re-interventions. Indeed, a significant influence was observed in the TGA group, where the number of reoperations correlates with lower vitality scores ($p = 0.009$).

Sixty-seven percent of the patients ($n = 102$) had undergone the last surgery before the age of two years, an age at which they are supposed to be not able to remember the experience of the operation. This parameter did not show any impact on the QoL ($p = 0.89$).

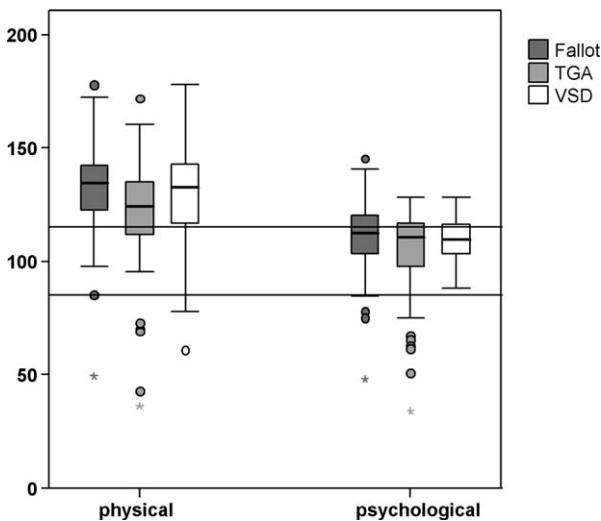


Fig. 2. Physical and psychological SF-36 scores assessed for each diagnosis group. No significant difference was revealed for physical and psychological aspects of the SF-36 between the diagnosis groups ($p = 0.12$).

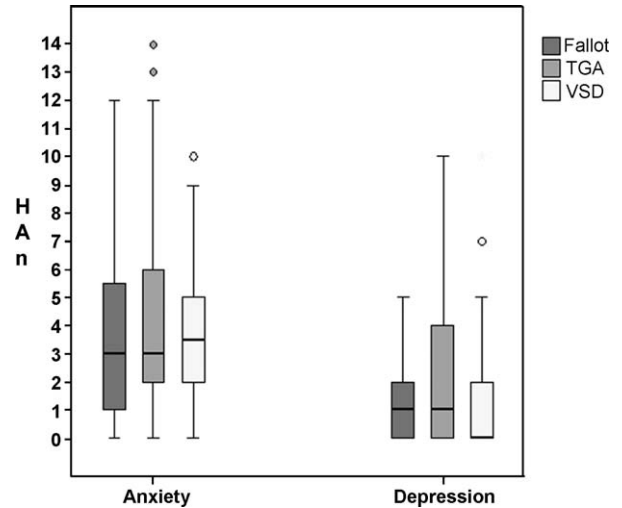


Fig. 3. HADS scores assessed for each diagnosis group. No significant difference was revealed for anxiety ($p = 0.32$) and depression ($p = 0.17$) scores between the diagnosis groups.

In contrast, the QoL seemed to be influenced by the time span between the last operation and the time of answering the questionnaires. In fact, the more time has passed, the lower the scores of the overall QoL ($p = 0.02$), in the physical ($p = 0.05$) as well as in the psychological ($p = 0.03$) aspects, most pronounced in the scales regarding general health ($p = 0.02$) vitality ($p = 0.003$) and depression ($p = 0.005$) (Fig. 5). This result was found throughout the whole sample, but most prominent among the TGA patients.

Concerning the NYHA functional class, the majority of patients (82%) were found to be in NYHA class I, 16% in class II, only 2% in class III, and none in class IV.

However, significantly more patients of the TGA group (47%), but only 23% of the TOF and 14% of the VSD group ($p = 0.001$) admitted to be limited by their cardiac malformation, mostly in regard to physical activities. Nevertheless, 76% of the patients declared that they perform sports and 83% declared that they do not consider that their QoL is limited by their malformation.

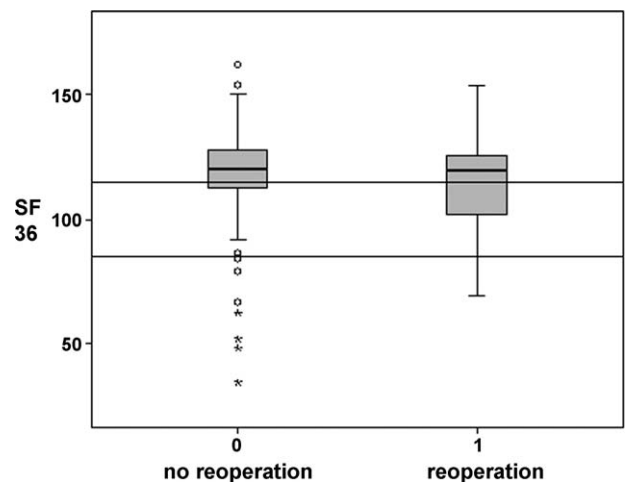


Fig. 4. Impact of reoperation on QoL. Reoperated patients showed no significant lower SF-36 scores ($p = 0.21$).

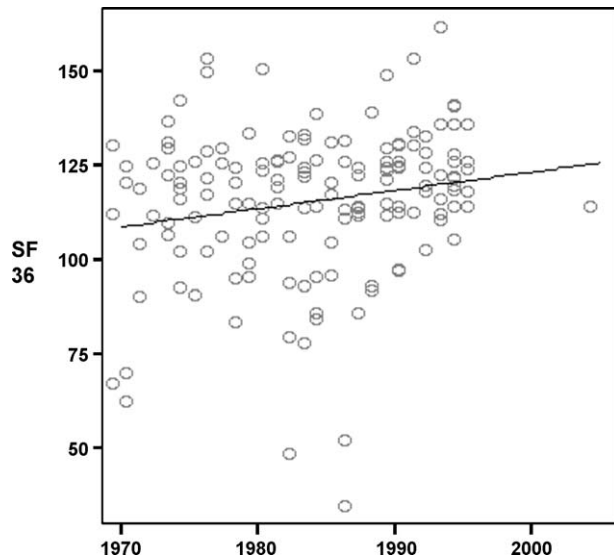


Fig. 5. Influence of the time span since the last operation. Analysis revealed that the more time has passed since the last operation, the lower were the scores for QoL.

Significantly more patients with TOF (93%) or TGA (95%) had regular consultation through a cardiologist compared to VSD patients (52%) ($p = 0.0001$). Evaluating long-term complications, arrhythmia were found to occur significantly more frequently within the TGA group (35%) than in the TOF and VSD group (8%) ($p = 0.0001$); however it was found to show no significant impact on QoL ($p = 0.10$). Anti-arrhythmic therapy was necessary in a significantly higher number of patients (26%) than in TOF (15%) and VSD (6%) patients ($p = 0.02$).

Of the 109 patients, who were older than 16 years and answered the question correctly, 76% patients ($n = 83$) declared having a fulltime job. In regard to disability pensions, 18% of the patients receive a full or partial pension, the majority of these among the VSD patients (22%), while only 18% of the TGA and 15% of the TOF patients did so, but without significant differences between the three groups ($p = 0.69$).

Problems with insurances were reported by 21% of the patients, most of them regarding health insurances (67%).

Regarding pregnancy and children, 4.4% of adult patients ($n = 5$, three women, two men), of whom four TGA and one TOF patients, declared to have renounced the idea of having children due to their cardiac malformation.

4.2. Analysis of the cyanotic versus acyanotic patients

Analysis revealed no lower QoL ($p = 0.65$) for the cyanotic group (TGA, TOF) and no higher anxiety ($p = 0.69$) nor depression ($p = 0.14$) scores compared to the acyanotic ones (VSD).

A further comparison between TGA patients after an atrial switch (Senning/Mustard, $n = 45$) versus an arterial switch ($n = 14$) operation, revealed, that GUCH after Senning or Mustard operation show significantly lower scores for QoL ($p = 0.03$), vitality ($p = 0.002$), psychological functioning ($p = 0.01$) and higher depression scores ($p = 0.02$) (HADS) compared to patients treated by the switch procedure (Fig. 6).

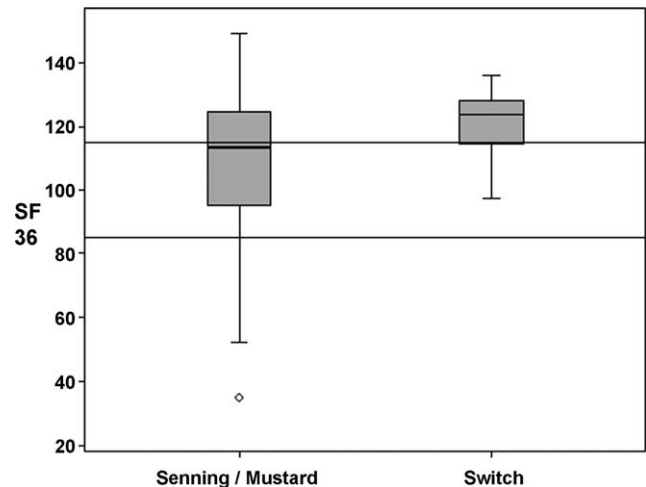


Fig. 6. Comparison of QoL following Senning/Mustard versus switch operation. GUCH patients after Senning/Mustard operation showed significant lower scores for QoL when compared to patients after switch procedure ($p < 0.01$).

5. Discussion

The assessment of the health related QoL is well established as a subjective indicator of health and to measure the benefits of therapy. It is evident, that health is a constituent of well being, but is not taken to be all that matters for well-being and for a good quality of life. Indeed, living with congenital heart disease affects patients not only physically, but also emotionally and socially. Frequently, it is assumed that patients with CHD have a reduced QoL, especially those who had undergone palliative surgical therapy.

Examining the QoL of adults with CHD contributes to the understanding of the impact of these conditions and to develop appropriate therapeutic concepts for these patients. In order to differentiate better the concept of 'QoL', the questionnaires were chosen according to a disease specific category (additional questionnaire) and to an overall category, so-called generic instruments (SF-36 and HADS) [5].

Surprisingly, the overall QoL of the whole collective was excellent, sometimes even better than of the age- and gender-matched standard population, without a significant difference between the three diagnosis groups. These findings differ from the results of a study by Lane et al. who assessed the QoL of 276 GUCH patients using the SF-36. They conclude, that physical functioning and overall general health perception was significantly lower in all CHD groups [7]. An explanation for this might be due to the aspect, that the authors did not transform the raw SF-36 values, allowing age- and gender-matched comparison and just compared the results to a standard population with an age between 30 and 34 years. In contrary to our expectation, the scores of the HADS test of the GUCH patients did not reveal higher levels of anxiety or depression. Assuming that these patients are exposed to an unusual level of anxiety about their future, their prognosis, and the possibility that their condition could get worse, these results are unexpected. The fact that these patients may feel somehow grateful and lucky to be alive

certainly influences positively their psychological attitude. However, they are exposed to particular psychosocial situations and problems, which requires support and should not be underestimated.

Age is intuitively and logically associated with a reduction of one's physical and mental abilities, which may impair the quality of life. Besides the scores of the SF-36, which are automatically corrected for age and gender, a separate analysis was performed and revealed that, age does not correlate with an impairment of the QoL of the overall collective. An explanation might provide the young age of the overall collective with a mean age of only 26 ± 11 years, a sample, which therefore might be too young to show a negative impact of age on their QoL.

Nevertheless, TGA patients appear more sensitive to this factor, because age significantly effects their general health, vitality, psychological functioning, and overall QoL, as well as their anxiety and depression scores. Even if 82% of these patients considered themselves as asymptomatic, and even if we could not show any other independent factors impairing their QoL, a progressive deterioration of their general functioning over time, perhaps related to their chronic right ventricular dysfunction, has to be assumed.

In contrast, the amount of time that had passed since the last operation was found to correlate with lower scores of the overall QoL, in both the physical as well as in the psychological aspects. This pattern of results was found in the scales regarding general health, vitality and depression, throughout the whole sample, and was even more noticeable in the TGA group.

A contributing factor is certainly the considerable improvement of the perioperative management, treatment algorithms, surgical techniques and standardization of follow-up over the last decades. Another potential explanation is suggested by Lane et al. proposing that childhood trauma together with the 'wrapped in cotton wool' attitude, which prevailed 20 years ago, could have contributed to a reduced QoL perceived by now adult patients [7]. However, the study by Lane et al. did not specifically examine the influence of the time span since the last operation on the QoL.

This underlines that QoL depends on a variety of factors, and psychosocial adjustment to adult life depends not only on the type and severity of the congenital heart defect, but also on the attitude and support of family, friends, society, and the medical care team. Sensitive handling, advocacy and education are enormously valuable.

The continuous care of GUCH patients should be provided through life. Indeed, most of these patients are at risk of complications arising from their initial lesion, treatment or repeated surgical procedures, and are at risk of premature death [3]. An important step is the transfer from pediatric to adult care. Our own experience shows that a narrow collaboration of pediatric and adult cardiologists is essential to minimize the number of patients, who are misplaced from continuous care and follow-up during this key period.

The guidelines of the task force for the management of GUCH patients of the European Society of Cardiology recommend a stratification of GUCH patients according to their risk level (low-, moderate- and high-risk), with a subsequent follow-up into three categories: patients requir-

ing (1) exclusively a specialized center (2) a general cardiac service or (3) non-specialized clinics [8]. In our sample, significant differences of follow-up were registered according to the diagnosis groups, with the majority of TOF and TGA patients, but only half of VSD patients, regularly visiting a cardiologist. This correlates well with the level of the recommended follow-up: TOF belonging to level 1 or 2, TGA after atrial switch to level 1, and VSD without complication to level 3. However, even if the majority is followed by a cardiologist, it has to be emphasized that only a minority is followed by a GUCH cardiologist in a specialized center, underlining the lack of transfer from the pediatric cardiovascular care to the GUCH specialist.

A well-recognized late complication observed in the GUCH patients is arrhythmias, which represent a frequent cause of morbidity and mortality. Furthermore, they are the main reason for hospitalization and are often associated with a decreased exercise tolerance and QoL [8–10]. Surprisingly, we could not observe a significant impact on the QoL, besides 35% of the TGA and 8% of the TOF and VSD patients suffer from arrhythmias. This is in accordance with the findings of a study by Irtel et al. who found arrhythmias in 25% of TGA patients and in 11% of TOF patients [9].

In regard to the relationship between the physical status and QoL, the self-assessment of the physical performance showed that 82% of the patients considered themselves as asymptomatic. In fact, 76% declared participation in sports as well as regular physical exercise. In addition, they all showed normal scores in the four aspects of the physical dimension of the SF-36 (physical functioning, role limitations caused by physical health problems, bodily pain, general health).

However, when the patients were asked whether or not they feel limited by their malformation in their daily life, 29% answered yes, and of the latter almost all of them confessed to feeling limited by a decreased exercise tolerance. Indeed, while many patients are able to enjoy the full range of normal life activities, patient symptom reporting should be interpreted with caution, as they may not know any better. De Bleser et al. investigated the self-reported physical activities of the patients after palliative treatment for TGA, and concluded that the level of physical activities of these patients is nearly normal, except for sport activities [11]. This limitation seems to have an effect on their day-to-day functioning in such a way that these patients tend to prefer activities that they can comfortably perform. Thus, the GUCH patients seem to adapt their way of living and avoid certain type of exercise and sport that make them feel symptoms and weaknesses, which remind them of their disease.

Nevertheless, even if 29% feel limited in their physical performances of their daily life, 83% claim that they do not feel limited in their QoL by their cardiac malformation; this without significant differences between groups. Thus, the physical component does not appear to influence their QoL as strongly as we expected. In fact, Kamphuis et al. already demonstrated that relating physical indices to the dimensions of health related QoL and subjective health status show weak correlation and that variables such as cardiac failure or arrhythmia (which are strong indicators of the objective severity of the disease) do not correlate with the patient's health related QoL, as we have observed in our patient population [12].

A significant part of the GUCH patients require multiple reoperations after the initial intervention. Reoperations of these patients are challenging and are combined with a high rate of complications, and the risk/benefit ratio of any proposed surgical procedure is difficult to assess [13]. Interestingly, the overall QoL stays unchanged in all groups, irrespective of whether the patient had multiple cardiac surgeries or just one intervention. We found an impairment of the vitality scores by the number of reoperations for the TGA patients only.

Furthermore and in contrary to our expectation, remembering the last operation or not, does not affect significantly the QoL. Sixty-seven percent of the patients were operated before the age of two, at an age where remembering the trauma of the operation itself cannot be expected. Even so, these patients did not show better scores. The experience of the operation, which is usually considered as a traumatic event, does not seem to leave such a shocking and disabling mark.

We observed a higher rate of complications (reoperation, arrhythmias) in the cyanotic group. However, these patients did not show either a lower QoL or higher anxiety or depression scores. These findings differ from those of Lane et al. who categorized the patients as surgically cured, corrected, palliated or inoperable [7]. The authors found that patients with cyanotic CHD had significantly poorer physical functioning as well as a poorer QoL. This difference could be explained by the fact, that the analyzed cyanotic patients were only palliated or inoperable, thus suffering from a chronic cyanosis (oxygen saturation <90% at rest), contrary to our patients, which were surgically corrected or palliated during early childhood.

Patients with CHD are at high risk of neurological complications. Indeed, low cardiac output, acidosis and hypoxia as well as the consequences of the cardiac surgery itself may affect their intellectual capacities. Furthermore, reduced physical capacity and absenteeism may make further education difficult and give rise to discrimination regarding job prospects. Eighteen percent of our collective said they received a partial or total disability pension without significant difference between the diagnosis groups. Surprisingly, we found the majority of these among the VSD patients despite the fact that cyanosis is often associated with a lower I.Q. average, higher rate of late complications, and a worse physical capacities [14,15]. This finding contrasts with the results of the study by Immer et al. who found a majority of GUCH patients on disability pension in the cyanotic group. But it has to be clarified that the categorization of the patient collective was rather broad and less targeted (ASD, outflow tract lesion, coarctation, and CAVC) and included patients with additional chromosomal aberrations [16].

Life and health insurance availability vary greatly both within and between countries. In Switzerland, health insurance is obligatory. Even so, 21% of the patients reported problems with insurances, most of them regarding health insurances (67%), without major differences between the three diagnoses. Costs of medical treatments are of great importance to market-depending and profit-orientated private health insurance companies, and in order to avoid the negative impact of financial considerations on the

provision of optimal medical care to GUCH patients, especially to those with worse outcomes, strategies need to be developed by the health care system, physicians, and organized patients groups to provide advice and assistance, also juridical if necessary.

Most GUCH patients can tolerate a pregnancy with appropriate care, but pregnancy counseling and complete evaluation is obligatory [17,18]. The impact of pregnancy on long-term survival and the risks to transmit the congenital heart disease to the fetus must be seriously discussed with the GUCH patients. Actually, 4.4% of our adult patients (three women, two men), of whom four TGA and one TOF patients, renounced the idea of having children due to their cardiac malformation. Indeed, patients with cyanotic heart disease are high-risk patients regarding pregnancy and should be managed by a specialized unit to guarantee multidisciplinary proper care.

6. Conclusion

This study shows that GUCH patients after congenital heart surgery have an excellent QoL, without any significant difference between diagnosis groups (TOF, TGA and VSD) or between the possibility of surgery (cure, correction or palliation).

Interestingly, analysis of potential negative factors, like age, number of reoperations, or arrhythmias did not show any significant impact on the QoL of the overall patient population.

However, TGA patients, of whom the majority had undergone an atrial switch procedure, were found to have a higher rate of complications and re-interventions, and appear to be more sensitive to age and to the time passed since the last operation, showing lower scores of QoL.

Although GUCH patients have an excellent QoL, they appear to be exposed to special psychosocial problems. Thus, a significant portion of GUCH patients receive a disability pension, report problems with life and health insurances, and even renounce having children due to their cardiac malformation.

These findings highlight the great importance of a structured medical follow-up provided by specialists for an adequate and complete management of these complex patients. The diverse needs of GUCH patients require multidisciplinary care, including psychologists, neurologists, mental health counselors, social workers, vocational counselors, obstetrics, genetics, and nurse specialists; ideally coordinated and provided in a GUCH center.

References

- [1] MacMahon B, McKeown T, Record RG. The incidence and life expectation of children with congenital heart disease. *Br Heart J* 1953;15(April (2)):121–9.
- [2] Sommerville J. Management of adult with congenital heart disease: an increasing problem. *Annu Rev Med* 1997;48:283–93.
- [3] Webb GD. Care of adults with congenital heart disease — a challenge for the new millennium. *Thorac Cardiovasc Surg* 2001;49(1):30–4.
- [4] Brazier JE, Harper R, Jones NM, O’Cathain A, Thomas KJ, Usherwood T, Westlake L. Validating the SF-36 health survey questionnaire: new outcome measure for primary care. *BMJ* 1992;305(July (6846)):160–4.

- [5] Bullinger M, Kirchberger I. Der SF-36 Fragebogen zum Gesundheitszustand. Handanweisung. Hogrefe Verlag. Gottingen.
- [6] Snaith RP. The hospital anxiety and depression scale. *Health Qual Life Outcomes* 2003;1:29.
- [7] Lane DA, Lip GY, Millane TA. Quality of life in adults with congenital heart disease. *Heart* 2002;88(July (1)):71–5.
- [8] Deanfield J, Thaulow E, Warnes C, Webb G, Kolbel F, Hoffman A, Sorenson K, Kaemmer H, Thilen U, Bink-Boelkens M, Iserin L, Daliento L, Silove E, Redington A, Vouhe P, Priori S, Alonso MA, Blanc JJ, Budaj A, Cowie M, Deckers J, Fernandez Burgos E, Lekakis J, Lindahl B, Mazzotta G, Morais J, Oto A, Smiseth O, Trappe HJ, Klein W, Blömstrom-Lundqvist C, de Backer G, Hradec J, Mazzotta G, Parkhomenko A, Presbitero P, Torbicki A. Task Force on the Management of Grown Up Congenital Heart Disease, European Society of Cardiology; ESC Committee for Practice Guidelines Management of grown up congenital heart disease. *Eur Heart J* 2003;24(June (11)):1035–84.
- [9] Irtel TA, Vetter C, Stuber T, Kuemin A, Heimes T, Pfammater JP, Tüller D, Carrel T, Delacrétaz E. Impact of arrhythmias on health-related quality of life in adults with congenital cardiac disease. *Cardiol Young* 2005;15(December (6)):627–31.
- [10] Li W, Somerville J, Gibson DG, Henein MY. Effect of atrial flutter on exercise tolerance in patients with grown-up congenital heart (GUCH). *Am Heart J* 2002;144(July (1)):173–9.
- [11] De Bleser L, Budts W, Sluysmans T, De Wolf D, Massin M, Gewillig M, Suys B, Moons P. Self-reported physical activities in patients after the Mustard or Senning operation: comparison with healthy control subjects. *Eur J Cardiovasc Nurs* 2007;6(September (3)):247–51.
- [12] Kamphuis M, Ottenkamp J, Vliegen HW, Vogels T, Zwinderman KH, Kamphuis RP, Verloove-Vanhorick SP. Health related quality of life and health status in adult survivors with previously operated complex congenital heart disease. *Heart* 2002;87(April (4)):356–62.
- [13] Berdat PA, Immer F, Pfammater JP, Carrel T. Reoperations in adults with congenital heart disease: analysis of early outcome. *Int J Cardiol* 2004;93(February (2–3)):239–45.
- [14] Daliento L, Mapelli D, Volpe B. Measurement of cognitive outcome and quality of life in congenital heart disease. *Heart* 2006;92(April (4)):569–74.
- [15] Newburger JW, Silbert AR, Buckley LP, Fyler DC. Cognitive function and age at repair of transposition of the great arteries in children. *N Eng J Med* 1984;310:1495–9.
- [16] Immer FF, Althaus SM, Berdat PA, Saner H, Carrel TP. Quality of life and specific problems after cardiac surgery in adolescents and adults with congenital heart diseases. *Eur J Cardiovasc Prev Rehabil* 2005;12(April (2)):138–43.
- [17] Warnes CA. Congenital heart disease and pregnancy. In: Elkayam U, Gleicher N, editors. *Cardiac problems in pregnancy*. 2nd ed., New York: John Wiley and Associates; 1988.
- [18] Siu SC, Sermer M, Harrison DA, Grigoriadis E, Liu G, Sorensen S, Smallhorn JF, Farine D, Amankwah KS, Spears JC, Colman JM. Risk and predictors for pregnancy-related complications in women with heart disease. *Circulation* 1997;96:2789–94.

Appendix A. Conference discussion

Dr M. Wojtalik (Poznan, Poland): As we all know, the transposition treatment has changed in the last two decades or three decades and the long-term result is different in these two groups after atrial switch and arterial switch. What kind of treatment did your patients get?

Dr Loup: The majority of the patients, 45 of the 59 patients, are status after a Senning or Mustard procedure. There were only 14 GUCH patients after switch operation. So it is difficult to compare these two groups, because one group is much younger and smaller.

Dr A. Corno (Liverpool, United Kingdom): I'm quite sure you are aware that in any test evaluation of the quality of life there is a very important subjective component related to the expectation of the patients and their family. Just to make myself clear, if the patient and the family have been told by the family doctor or by the cardiologist that after heart surgery procedure they can forget about sports and so forth, the children are quiet, the teenagers are quite happy spending their life watching TV or playing Game Boy, like a normal teenager, by the way, and this creates a very low expectation. So if you ask if they are happy, of course they are happy because they have been told they cannot do much more. How did you control this subjective factor in your test?

Dr Loup: This is a very good point. One has to say that this is almost impossible to control. However, the tests are standardized and well evaluated in many studies and should consider such subjective factors.

Dr L.H. Edmunds (Philadelphia, Pennsylvania, USA): Well, a control would be a child that had a different operation, like an abdominal operation, rather than just a normal, the norm for the test, because you had a very wide standard deviation for all of those factors.

Dr O. Ghez (London, United Kingdom): Could you elaborate a bit more on the almost 200 patients that were not able to answer the questionnaire. Should we be worried?

Dr Loup: Yes, indeed, we should be worried. We were more than surprised after realizing how many patients were lost from follow-up. Unfortunately, this seems to be the reality and we are keen to change this together with our cardiologists.

Dr M. Hazekamp (Leiden, The Netherlands): It's especially interesting that you found in your transposition patients you had such a high... well not, let's say, not a very good, not a very perfect quality of life score. In the afternoon I will present our follow-up on arterial switch operations for transposition, with 20 years follow-up, and they did significantly better. So there must be some reason for that. And would it be only the number of reoperations, do you think, that changed your quality of life towards worse?

Dr Loup: I didn't understand the end of your question, I apologize.

Dr Hazekamp: Was the number of reoperations significantly higher in the Mustard and Senning population?

Dr Loup: Yes. The number of reoperations was significantly higher in the Senning/Mustard group. However, I think, that a longer follow-up of the switch patients is necessary in order to allow for a comparison of the two groups.

Dr Hazekamp: Yes, that would be nice.