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Original Article

Are cyanosed adults with congenital cardiac malformations depressed?

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Abstract *Objective:* To assess the incidence of depression, and the ability to interact socially, in adult patients with chronic cyanosis and congenital cardiac malformations. *Design:* Prospective study of consecutive patients. *Setting:* Single institution, tertiary referral centre. *Patients:* Between 1993 and 2000, we assessed 76 patients with congenital cardiac malformations and persistent cyanosis, having a median age of 36.5 years, with a range from 19 to 64 years, at the time of referral. Female patients accounted for just under half (48.6%) of the sample. Just under two-fifths of the cohort (39.5%) had functionally univentricular cardiac anatomy, while 14.8% had tetralogy of Fallot with pulmonary atresia and aorto-pulmonary collateral arteries, and 17% had the Eisenmenger syndrome. During the period of follow-up, 17 (22.4%) of the patients died. *Assessment:* We used clinical interviews and non-invasive assessment, employing Zung's questionnaire which provides a scale for the self-rating of depression. On this scale, a score above 50 points is indicative of depression. *Results:* Of the survivors, 32 (54%) completed the self-rating questionnaires. Of these, 20 responders (63%) considered that they lead full lives, including sexual activities, while 26 (81%) had never harboured suicidal thoughts. Depression was diagnosed in 11 responders (34%), with a mean score of 66.9, standard deviation of 8.7, and a range from 53 to 89. The remaining 21 patients (66%) were without signs of depression, scoring a mean of 41.5, with standard deviation of 5.5, and a range from 35 to 46. Depression was associated with older age (40.5 years versus 33.5 years, $p = 0.01$), worse functional state in the classification of the New York Heart Association (2.95 versus 2.48, $p = 0.03$), and unemployment ($p < 0.0001$), but independent from the severity of cyanosis, the level of the haematocrit, the saturation of oxygen, or previous surgical treatment. *Conclusions:* To our knowledge, this is the first evidence suggesting a relatively high incidence of depression in adults with congenital cardiac malformations and persistent cyanosis. Larger, multi-centric studies will be needed to confirm or refute these findings.

Keywords: Congenital heart defects; psychosocial aspects; Zung's self-rating depression scale

The term "inoperable" seems inappropriate nowadays when almost every patient with a congenitally malformed heart can undergo surgery, or at least considered for transplantation of the heart, or the heart and lungs. For many patients presenting during the 1950's and 1960's, nonetheless, simple palliation was the only, and then definitive, solution for their cyanotic congenital cardiac disease. In this setting of complex congenital cardiac lesions, patients with a well-balanced circulatory pattern were often left without any further surgery. To this group of chronically cyanosed patients must then be added those with the Eisenmenger syndrome. The long-lasting tissue hypoxia which accompanies this cyanosis is known to have many adverse consequences on various systems in the human body.[1]

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The new centres for adolescents and adults with congenital cardiac disease are supposed to provide the advice, support, and the best available treatment needed by these patients. Decision-making is often difficult in this setting of chronic cyanosis because of scarcity of information relating to their long-term management and outcome.[2–6] There is very little doubt that any further surgical treatment would carry great risks of early and medium-term postoperative complications, and even death, when compared with similar malformations as currently seen in children. Furthermore, there are important social issues, such as support of the family, children, partners, employment, and so on, to take into account. The decision is further complicated by the fact that the adults with congenital cardiac disease and persistent cyanosis are usually well adapted to hypoxia and their rather restricted way of life.

The aim of this study, therefore, was to assess the perception of well-being, and the ability to interact socially, in a cohort of adults with chronic cyanosis due either to complex congenital cardiac malformations or the Eisenmenger syndrome.

Patients and methods

Between January 1993 and August 2000, 76 patients with congenital cardiac disease and persistent cyanosis, having a saturation of oxygen less than 90%, had been referred to and assessed in the Centre for Adults with Congenital Heart Defects, University Hospital Motol, Prague, Czech Republic. Their median age was 36.5 years, with a range from 19 to 64 years, at the time of referral, with female patients representing 48.6% of the cohort. In terms of anatomic diagnoses, 30 patients (39.5%) had functionally univentricular cardiac anatomy, 11 patients (14.8%) had tetralogy of Fallot with pulmonary atresia and major aorto-pulmonary collateral arteries, and 9 patients (11.8%) had tetralogy of Fallot with pulmonary stenosis but either without previous surgery, or else with only palliative surgery in the past. In addition, 13 patients had Eisenmenger syndrome (17%), 6 patients were seen with unoperated Ebstein's malformation of the tricuspid valve (7.9%), 3 patients had complete transposition with atrial and/or ventricular septal defects, 3 patients had congenitally corrected transposition with pulmonary stenosis and atrial or ventricular septal defect, and one patient had untreated totally anomalous pulmonary venous connection (Table 1). Of the patients, 27 (35.5%) had undergone 35 previous palliative surgical procedures at an interval of 25 ± 9.1 years, the procedures having been performed between 1952 and 1993 (Table 2).

Table 1. Adults with congenital cardiac disease and persistent cyanosis.

Diagnosis	N	%
Functionally univentricular heart	30	39.5
Eisenmenger syndrome	13	17
Tetralogy of Fallot with pulmonary atresia and multiple aorto-pulmonary collateral arteries	11	14.8
Tetralogy of Fallot with pulmonary stenosis	9	11.8
Ebstein's malformation	6	7.9
Complete transposition with ventricular and/or atrial septal defect	3	4
Congenitally corrected transposition with pulmonary stenosis and ventricular and/or atrial septal defect	3	4
Totally anomalous pulmonary venous connection	1	1
Total	76	100

Table 2. Previous palliative surgical treatment.

Type of surgery	N	Period
Blalock-Taussig shunt	21	1952–1985
Classical Glenn shunt	7	1964–1981
Aorto-pulmonary shunts	3	1972–1990
Bidirectional cavopulmonary anastomosis	1	1993
Total cavopulmonary connection	2	1990–1993
Banding of pulmonary trunk	1	1972
Total	35	

All referred patients were assessed clinically, and further investigated using an electrocardiogram, transthoracic and/or transoesophageal echocardiography, full blood count, haemocoagulation tests, and biochemical analysis. In 15 patients, the decision was made to undertake further surgical treatment (Table 3).

Zung's questionnaire, which permits depression to be self-rated on a formal scale,[7] was completed by 32 patients out of 59 survivors (54%). The diagnoses, age at assessment, values for saturation of oxygen, haemoglobin, haematocrit and previous surgical treatment for these patients are listed in Table 4.

Statistical methods

We used Student's t-test for analysis of normally distributed quantitative variables, and the chi-square test for qualitative variables. A P-value of less than 0.05 was considered significant.

Results

Of the cyanotic patients referred for assessment, 26 (34.2%) were in the second class of the grading system developed by the New York Heart Association,

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Table 3. New surgical treatment.

Patient	Born	Diagnosis	Previous surgery/year	New surgery/year
MD	1965	TA	BT/67	TCPC/94
JP	1959	TA	Glenn/64	TCPC/96+
TM	1974	TA	BT/75, Glenn/81	BT/99
JN	1939	TOF	0	ICR/95+
MF	1966	TOF	0	ICR/96
PO	1971	CTGA, VSD, PS	Pulmonary banding/72	ICR/99
OS	1953	TAPVC	0	ICR/96
IR	1964	DILV, PS	BT/79, Wat	exc.m./95
JK	1968	DILV, PS	BT/78, BCA/93	TCPC/96
TE	1975	DORV, VSD, PS	BT/78	ICR/95+
EB	1972	FSV, AVSD, PS	BT/75, Glenn/78	APA/99
MR	1962	CTGA, PS, ASD	0	ICR/00
PC	1965	FSV, PS	BT/77	TCPC/00
MS	1960	DILV, PS	BT/66	APA/93
MP	1953	criss/cross, DORV, PS, ASD, VSD	0	BT/00

Abbreviations: TA:tricuspid atresia, TOF: tetralogy of Fallot, CTGA: congenitally corrected transposition, VSD: ventricular septal defect, PS: pulmonary stenosis, TAPVC: totally anomalous pulmonary venous connection, DILV: double inlet left ventricle, DORV: double outlet right ventricle, FSV: functionally single ventricle, AVSD: atrioventricular septal defect, criss/cross: criss-cross heart, ASD: atrial septal defect, BT: Blalock-Taussig shunt, Wat: Waterston shunt, BCA: bi-directional cavo-pulmonary anastomosis, APA: aorto-pulmonary anastomosis.

Table 4A. Data on patients without depression.

Age (years)	NYHA	Em	Ed	chil	Hb (g/l)	Sat (%)	HTC	Score	dg	Previous surgery	New
30	2.5	N	Y	1	180	70	54	46	EIS	0	0
48	3	N	N	3	156	81	52	43	EIS	0	0
22	2	Y	Y	0	210	91	63	41	EIS	0	0
60	4	N	Y	1	171	81	51	45	EIS	0	0
35	2	Y	Y	0	165	80	49	49	TA	BT	TCPC
27	2.5	Y	N	0	226	81	68	46	TA	BT, GLENN, APA	0
26	2	Y	Y	0	239	77	73	41	TA	BT	BT
24	2.5	Y	Y	0	156	81	46	49	PA	0	0
24	3	Y	N	0	172	82	52	49	PA	BT	0
24	2	Y	Y	0	182	80	52	35	PA	BT	0
28	2	Y	Y	1	171	72	53	35	MA	BT	0
34	2	Y	Y	0	185	83	57	39	FSV, DORV	0	0
26	3	Y	N	0	183	87	56	38	DORV	BT	0
38	2	Y	Y	0	203	85	57	38	FSV, DORV	0	0
32	2	Y	Y	2	197	91	59	38	DILV	BT, BCPA	TCPC
35	3	Y	Y	0	169	75	51	43	FSV	BT	TCPC
28	3	Y	Y	0	208	79	63	41	FSV	BT, GLENN	AO-AP
42	2	Y	N	0	163	78	51	41	DORV	0	0
44	3	Y	N	0	191	76	61	46	EIS	0	0
22	2	N	N	0	185	88	66	45	DORV	TCPC	0
55	2.5	Y	Y	2	155	83	54	24	TA	BT	0
33.5	**2.4**	**71%**	**67%**	**1.7**	**184**	**81**	**57**	**41.5**		**57%**	**24%**
SD 8.782	0.4			28%	16	4.9	4.4	5.5			

35 were in the third class (46.1%), and the remaining 15 patients (19.7%) were rated in the fourth class. The mean saturation of oxygen at rest was 78.8%, with a standard deviation of 12%, and a range from 47 to 90%. The mean values of haemoglobin and haematocrit were 179 g/l (SD 22.8 g/l, range 120–239 g/l) and 55% (SD 6.7%, range 37%–73%), respectively.

Since 1993, 17 patients (22.3 %) have died during follow-up at a median age of 29.9 years, and with a range from 19 to 56 years. Of the deaths, 7 were sudden (41%), while 5 patients (29%) died due

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Table 4B. Data on patients with depression.

Age	NYHA	Em	Ed	chil	Hb (g/l)	Sat(%)	HTC	Score	dg	Previous surgery	New
28	2.5	N	N	0	180	50	54	89	PA	BT	0
36	4	N	N	0	218	74	58	75	TOF	0	0
36	3.5	N	N	0	216	77	51	63	FSV	BT	ex.m.
44	2.5	N	Y	2	175	85	52	69	FSV	0	0
47	3	N	N	0	161	54	56	68	DORV	0	BT
38	3	N	N	0	206	90	58	63	CTGA, ASD	0	ICR
50	2	N	N	2	151	83	49	53	TOF	GLENN, BT	0
40	3	N	Y	1	190	69	60	56	FSV	BT	APA
31	3	N	N	3	143	75	49	68	PA	0	0
45	3	N	N	3	198	89	60	69	EIS	0	0
51	3	N	N	2	168	89	52	63	DORV	BT	0
40.5	**2.9**	**0%**	**18%**	**2.2**	**182**	**76**	**54**	**66.9**		**45%**	**27%**
SD 6.9	0.4			54%	23	13	3.8	8.77			
p: 0.010	0.03	0.00	0.02	0.29	0.6	0.2	0.3	0.000		0.79	0.83

Abbreviations: TA: tricuspid atresia, PA: pulmonary atresia, EIS: Eisenmenger syndrome, DORV: double outlet right ventricle, CTGA: congenitally corrected transposition, FSV: functionally single ventricle, ASD: atrial septal defect, TOF: tetralogy of Fallot, BT: Blalock-Taussig shunt, APA: aortopulmonary anastomosis, ICR: intracardiac repair, TCPC: total cavopulmonary connection, ex.m.: excision of supramitral membraine, Em: employment, Ed: higher education, chil: children Y: yes, N: no, HTC: hematocrit.

to congestive heart failure complicated by the low cardiac output. Three patients died following new cardiac surgical procedures giving a postoperative surgical mortality of 20%. No details about the cause of death were available in the remaining 2 patients.

Of the survivors, 95% were self-sufficient, with 18 patients (24%) in part or full-time employment. Higher levels of education had been achieved by 16 patients (21%), and 13 patients (17%) had either their own or adoptive children.

From this group of survivors, 32 completed the questionnaire permitting self-assessment of their levels of depression. Of these, 20 (62.5%) declared that they lead a full life, including satisfactory sexual activities, while 26 (81.3%) had never had suicidal thoughts.

In contrast, as based on Zung's questionnaire, the scores achieved by 11 of the 32 responders (34%) pointed to the presence of depression. The mean score of this group was 66.9, with a standard deviation of 8.7, and a range from 53 to 89. The scores from the remaining 21 patients (66%) did not indicate any signs of depression, the mean being 41.5, with a standard deviation of 5.5, and a range from 35 to 46. This was reflected in the higher number of employed patients in the group without depression (17 of 21 patients – 71%) when compared with the patients with depression. All of the latter group were without employment, two of them having given up work for health-related problems 2 and 26 years ago ($p < 0.0001$). The level of education was significantly higher in the group without depression (67% had higher education, $p = 0.026$). A higher grading in the classification of the New York Heart Association was also found in the patients with depression (mean 2.95, SD 0.48) when compared with their happier counterparts (mean 2.48, SD 0.45, $p = 0.03$). The group without depression was also younger (33.6 years, SD 8.7 vs. 40.5 years, SD 6.8, $p = 0.01$). No differences were found, however, in the saturations of oxygen ($p = 0.2$), the levels of haemoglobin ($p = 0.5$), and the haematocrit ($p = 0.3$). There was also no difference discovered in the number of previous palliative surgical procedures (57% versus 45%, $p = 0.79$) between those without and with signs of depression (Table 4).

Discussion

Adolescents and adults with congenital cardiac malformations and persistent cyanosis represent a challenging group of patients. Those with the Eisenmenger syndrome form part of this group, and it is well known that these patients may survive till middle age, although their mortality is high, with one-third dying during a 7 year period of follow-up in one study.[2] The natural history of the patients with functionally univentricular hearts is also associated with a high mortality (36%, even in the younger age, particularly in those without concomitant pulmonary stenosis.[6] The diagnosis of Eisenmenger syndrome, or complex congenital cardiac disease, still remains a significant risk factor for an adverse outcome to transplantation of the lungs or heart and lungs, particularly in the older patients.[3,8] The disappointing long-term results thus far of Fontan, or Fontan-like,

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operations[9,10] may strengthen the case for alternative long-term surgical palliative procedures.[4,5]

Moreover, there is likely to be a significant perioperative and early postoperative mortality related to any contemplated cardiac surgical treatment in the adults with complex congenital cardiac defects. Our experience, with 20% surgical mortality, corresponds with the experience of Dittrich et al.,[11] who found 16% mortality. All the above factors make the decision about undertaking any form of further surgical treatment very difficult.

The future well-being of each individual patient will then depend not only on the best advice and effort related to the medical and surgical treatment, but also on the perception of his or her current quality of life made by the patient. Our results indicate that, although most of these patients are likely to be self-sufficient, with 95% of our cohort achieving this situation, the rate of depressive scoring (34%) in a smaller subgroup of patients is alarming. This incidence of depression appears to be strongly linked with unemployment, a lower level of education, and older age, all of these findings being in agreement with the initial report of Zung and colleagues.[12] We found no correlation between signs of depression and either saturations of oxygen or the levels of haemoglobin.

Although the originally over-protective and anxious family environment[13] could have played its role in the ability of the patients to gain the employment later on, some reluctance of the employers to engage these chronically ill patients cannot be excluded. Unfortunately, we did not assess the efforts of the patients actively to gain employment. Although those patients with depression were graded at a higher point in the functional class of the New York Heart Association, we believe that the lack of social contact, and failed aspirations related to unemployment, were the main reasons leading to their depression. Our results complement the findings of Gupta et al.[13] They found a similar incidence of depression using a different technique to assess children with cyanotic congenital malformations, and uncovered depression in one quarter of their small cohort of 24 patients. This trend could have been, among other things, related to the reported increased maternal anxiety surrounding these patients. A similar mechanism could have lead to our findings of lower educational achievements in our depressed patients.

Most of our patients were physically well adapted to their hypoxaemia, albeit that tissue hypoxia lasting for decades could have deleterious effect on their psychological well-being, as shown by the higher incidence of depression in the older patients. As we used only Zung's questionnaire to assess depression in our patients, we could not fully exclude the influence of somatic or psychosomatic problems on the final score. A comparable control group from the same geographic area is currently not available.

Depression is often unrecognised,[14,15] and the data concerning its prevalence are variable. Depression is estimated to affect 2–4% of those in the community, 5–10% of patients in primary, and 10–14% of medical inpatients.[16] In studies using Zung's Self-Rating scale, the prevalence of depression has ranged from 6 to 20.9%.[2,14,15,17,18] In patients assessed one year after myocardial infarction using this questionnaire, one-third have been shown to be depressed, as in our study, and the clinical features of depression were the same as in patients without any history of cardiac disease.[19] In patients with stable congestive cardiac failure and coronary arterial disease, the depressed mood was significantly related to increased mortality.[20,21]

Although psychological problems are common in young adults with chronic illness,[22] including those with congenital cardiac disease,[23] there is an acute need for the detailed psychological assessment of all the adults with cyanotic malformations, as they seem at a higher risk of depression. Psychological support, or even psychiatric treatment, should be made available to those in need.

To our knowledge, this is the first report on the incidence of depression in a relatively small group of adults with cyanotic congenital heart defects. A potential weakness of our study is that only half of our group completed the questionnaire to assess their propensity to depression. Thus, larger multi-institutional studies are urgently needed to confirm or refute our findings. The results would allow for the appropriate planning and allocation of resources in the care of this steadily growing group of patients.

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