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Penelitian Klinik

Surgical Risk for Adult with Tetralogy of Fallot

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Background. The beneficiaries of successful pediatric cardiac surgery and cardiology increases surgical treatment in the adult population with congenital heart disease. Adults patient with Tetralogy of Fallot (TOF) will need surgery either reoperative, palliative or more commonly reparative by the time they come. Many of them face the prospect of further operations, arrhythmias, complications and if managed inappropriately will increase risk of heart failure and premature death. This study presents the early surgical results of adult TOF.
Methods and results. Data were reviewed retrospectively related to the hospital course of TOF adult patients (age $\geq$ 17 years) from June 1, 1998 to December 30, 2006 who require surgical treatment in National Cardiac Center - Harapan Kita, Indonesia. Most of the operations were corrective procedures (20 patients, 66.7%), followed by palliative procedures (6 patients, 20%), and reoperations (4 patients, 13.3%). Mean age at surgery was 20.87$\pm$5.49 years. There were no hospital mortality in the primary corrective group, one death (16.7) in the palliative group related to poor left ventricular function, and one death (25%) in the reoperation group related to perioperative bleeding. Follow up data of 11.73$\pm$17.68 months duration were available in 28 of 30 patients (93%) who were discharged home. Overall survival probability is higher for corrective procedures than palliative and reoperative procedures.
Conclusions. Surgical treatment of TOF in adult patients proved quite save and beneficial

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Keywords: Adult Tetralogy of Fallot, palliative procedures, corrective procedures, reoperations.

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Clinical Research

Risiko Bedah pada Pasien Dewasa dengan Tetralogi Fallot

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Latar Belakang. Keberhasilan bedah jantung anak telah membawa manfaat besar bagi populasi dewasa dengan penyakit jatung bawaan. Pasien dewasa dengan Tetralogi Fallot (TF) akan memerlukan tindakan bedah, mungkin reoperasi, bedah paliatif atau yang paling sering bedah reparatif/korektif. Bila tidak ditangani dengan baik, kebanyakan pasien ini akan menghadapi masalah bedah berulang, aritmia, gagal jantung dan kematian dini. Penelitian ini mempresentasikan hasil dini bedah pada pasien dewasa dengan TF.

Metode dan hasil. Data rekam medis pasien dewasa dengan TF (usia 17 tahun atau lebih) yang menjalani pembedahan di Pusat jantung Nasional – Harapan Kita, Jakarta, Indonesia dalam periode 1 Juni 1998 sampai dengan 30 Desember 2006 dipelajari secara retrospektif. Sebagian terbesar telah dilakukan bedah korektif (20 pasien, 68%), diikuti dengan bedah paliatif (6 pasien, 20%), dan reoperasi (4 pasien, 13.3%). Usia rerata saat bedah adalah 20.87±5.49 tahun. Tak ada kematian pada kelompok bedah korektif primer. Satu kematian terjadi pada 6 kasus yang menjalani bedah paliatif (16.7%), dan satu kematian (25%) terjadi pada 4 kasus yang menjalani reoperasi pasca bedah paliatif sebelumnya. Data evaluasi lebih lanjut didapat dari 28 pasien (93%) yang diikuti selama periode rerata 11.73±17.68 bulan. Probabilitas survival keseluruhan lebih tinggi pada kelompok bedah korektif dibanding kelompok paliatif dan reoperasi.

Kesimpulan. Pembedahan pada pasien dewasa dengan TF terbukti aman dan bermanfaat.

Kata Kunci: Tetralogi Fallot dewasa, bedah paliatif, bedah korektif, reoperasi.

The number of patients with adult congenital heart disease (ACHD) is expected to increase during this decade.[1] Surgery for ACHD as a result of primary repair of CHD within the pediatric age group is still challenging. Although the outlook for patients with CHD has been transformed, most early interventions have not been curative. It is estimated that approximately 50% of ACHD patients face the prospect of further surgery, arrhythmia, heart failure and - if managed inappropriately – of premature death.[2,3]

Surgery for Tetralogy of Fallot (TOF) as a benchmark of cyanotic type congenital cardiac surgery has been chosen to be analyzed. Adult patients with TOF will have had surgery either reoperative, palliative or more commonly reparative by the time they come. The objective of this study is to present the early surgical results of patients who require surgery for TOF in the adult population.

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Material and Methods

We reviewed the medical records of adult patients (aged 17 years or older) who underwent cardiac operations at

the National Cardiovascular Centre - Harapan Kita Hospital, Jakarta-indonesia, from June 1, 1998, through December 31, 2006.

Cardiac rhythm, preoperative risk factors, extracardiac anomalies, and postoperative complications were identified as risk factors. For analysis, surgical procedures were divided into three categories, including the following: (1) palliative procedures; (2) corrective procedures; and (3) reoperations. Palliative procedures include all operations performed to improve the patient's clinical status. Corrective procedures are operations employed to achieve an anatomical or physiologic correction with separation of the pulmonary from systemic circulation.

Results are presented as mean and standard deviation (median and range were used if data were not normally distributed). Comparisons of continuous variables were performed by Welch,s t-test. P value greater than 0.05 means that difference between groups is not statistically significant. To assess if there was a statistical significance between groups, data were analyzed with analysis of variance. Multiple comparison test was further employed to assess statistical significance between groups. Overall survival probability, as well as survival probability for different surgical categories (palliative procedures, corrective procedures, and reoperation), were analyzed according to Kaplan–Meier method.

Results

Thirty patients (≥17 years of age) requiring surgical treatment for TOF were included in this study which is consist of 13 males and 17 females. Mean age at surgery was 20.87±5.49 years. Extra cardiac anomalies were identified in 5 patients and other intracardiac anomalies were found in 4 patients. Preoperative diagnosis was achieved by echocardiography and cardiac catheterization.

According to preoperative clinical evaluation, 28 patients belonged to New York Heart Association (NYHA) class I-II, 2 in NYHA class III. Sinus rhythm in 20 of 30 patients (67%) was revealed in electrocardiogram. Arrhythmia were present in the remaining 10 patients and including Right Bundle Branch Block (RBBB) 7 patients, Ventricular Extra Systole (VES) 3 patients, sinus bradycardia 1 patient. Preoperative variables which are commonly associated to an increase hospital morbidity and mortality were identified in 7 patients, being multiple in 2 patients. Preoperative arrhythmia and preoperative NYHA class III-IV were the only causes linked to increasing hospital mortality.

Major postoperative complications occurred in 10 patients (30%). Seven patients presented multiple complications (**Table 1**). Pulmonary insufficiency and right ventricular dysfunction were measured by echocardiography following intracardiac repair, with 10 patients mild pulmonary stenosis (PS), 7 patients

Table 1. Postoperative Complications

Complication	n
Arrhythmias	6
Bleeding	3
Low output syndrome	2
Pericardial effusion requiring drainage	1
Pleural effusion	1
Cardiac arrest	1
Neurological deficit persisting at discharge	1
Other postoperative complication	1

Table 2. Surgical Categories

Variables Analyzed	Corrective Procedures	Palliative Procedures	Reoperations	p Value
Number of procedures	20(66.7%)	6 (20.0%)	4(13.3%)	
Sex M/F	5/15	6/0	2/2	0.05
Mean age in years (SD)	20.35(5.81)	21.8 (4.02)	22.0(6.78)	0.78
Preoperative sinus rhythm	12(60%)	4 (66.7%)	2(50%)	0.87
Postoperative complications	3(15%)	6 (100.0%)	1(25%)	0.01
Sinus rhythm at discharge	18(90%)	4 (66.7%)	4(100%)	0.24
Mean ICU stay/days (SD)	0.78(0.55)	1.22(1.71)	0.62(0.44)	0.49
Mean hospital stay/days (SD)	15.40(8.93)	9.67(6.97)	9.75(6.40)	0.23
Hospital mortality (%)	0(0%)	1(16.5%)	1(25%)	0.10

mild pulmonary insuffisiensi (PI) and 2 patients with severe PI. Two patients with moderate to severe right heart failure.

There were 2 hospital deaths (6.4%), one in the palliative surgery group and one in the reoperation group; no hospital death in the corrective group. The most common causes of death were pump failure and perioperative bleeding. Patients who underwent corrective procedures presented with lower mortality rate, requiring also less time in the intensive care unit and a shorter total hospital stay, although is was not significant (**Table 2**).

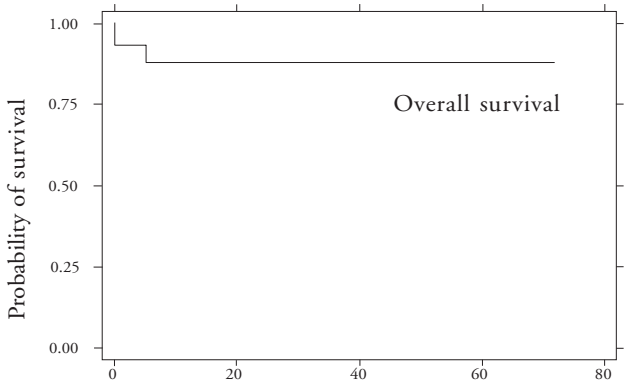

Figure 1. Overall survival probability according to Kaplan-Meier function with 95% confidence bands. Overall survival in 60 months probability is 87.84% with 95% confidence bands (0.66-0.96)

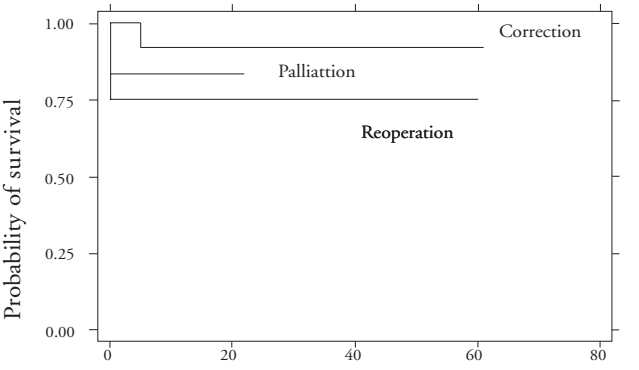

Figure 2. Kaplan-Meier survival function by type of surgery (palliations , corrections, reoperation). Survival in 60 months probability in total correction type is 92.31% with 95% confidence bands (0.57-0.99), in reoperation type is 75% with 95% confidence bands (0.13-0.96) and in palliation type is 83.33% with 95% confidence bands (0.27-0.97)

Follow up data were available in 28 of 30 patients (93%) who were discharge from hospital. Reoperation was necessary in 2 of 30 patients (6%) within a mean time 3 months after hospital discharge. At a mean follow-up time of 11.73$\pm$17.68 months 28 patients (93%) were in NYHA I-II. Late death occurred in 1 patient (3%). Survival probability according to Kaplan–Meier showed an overall survival of 87.8% at 60 months (**Figure 1**). A follow-up analysis shows an overall survival probability of 87.8% at 60 months, which is higher for corrective procedures (92.3%) if compared with reoperations (75%) and palliations (83.3%) (**Figure 2**).

The significant p value is postoperative complications ($p<0.05$), and there are no statistically significantly different in the multiple comparison (Bonferroni) test for mean comparison.

Discussion

Patients with CHD who reach the adult age usually remain in good or mildly decreased functional status for several years and are eventually referred to surgical treatment because of increasing symptoms not controlled by medical therapy. The majority of these patients survived into adult age in good functional status, considering that 93.3 % of them were preoperative NYHA class I or II. They are a selected group of patients mainly composed by individuals who have survived into adult age without irreversible damage to their heart and lung. Our data suggest that adult TOF patient with good functional capacity (NYHA class I-II), the hospital outcome can be better when compared to that of patients who presented with worse functional status (NYHA class III-IV).

The overall hospital mortality for TOF repairs in our centre during three years period (2003-2005, N=273) was 6.23%, comparable to the 5% mortality rate from the European Congenital Database. In our series, there is no hospital mortality for TOF repair in adult. Palliative procedure was offered to our adult TOF patient with poor left ventricular function, to improve the clinical condition at least temporarily. In our series there were 6 cases who had palliative procedure, with hospital mortality of 16.7% (1 patient). Although the operations achieved a decrease on cyanosis, we have no long-term functional results of these patients. Reoperation after a previous palliative procedure to enlarge pulmonary artery,

carried a high mortality rate (one out of 4 patients, 25%). The cause of death was related to perioperative bleeding. We have no reoperation related to conduit placement or pulmonary valve replacement after initial correction. The better outcome in the corrective group possibly related to the better condition of the patients.

Deterioration of RV function and exercise capacity correlate with a longer interval since repair, but these moderate associations can not be used to predict the pace of decline in the individual patients. Improvement of clinical condition may be obtained by improving right ventricle outflow tract (RVOT) function preservation during surgery, and by reducing the effect of electrical inhomogenicity for long term outcome of TOF.[5] The oldest patient in our series is a 42 years old patient with poor right ventricular function and arrhythmia, the long term outcome in this particular patient is also poor.

Optimal timing for reoperation pulmonary valve replacement seems crucial, as the children became an adult, the criteria so far have not been established. We have seen this problem in a 26 years of age patient with absent pulmonary valve. Shimazaki et.al[6] showed that in his series, beyond the age 30 to 40 progressively more patients become symptomatic, with onset of right heart failure.

A follow-up analysis shows an overall survival probability of 87.8% at 60 months, which is higher for corrective procedures (92.3%) if compared with reoperations (75%) and palliations (83.3%). Although palliative operations in these kinds of patients did achieve a decrease cyanosis at low risk, but we have no long-term functional results of these patients to allow definitive conclusions. Reoperation after a palliative procedures or previous corrective procedure includes patients with either a residual lesion after correction or patients who acquired secondary complications after their initial correction. These group of patients show the most difficult challenging, who need the premium hands of congenital adult cardiac surgeon. It is important to know the long term post-operative functional results of these patients. Unfortunately we were able to collect follow-up information only in 67% of the patients, and limited to 60 months after the operation (mean of 11.73 $\pm$ 17.68 months). The reason for this limitation is related to the patients social economy condition, most of them were referred from peripheral suburban area, no proper follow up could be performed.

Continuous evaluation of the quality of care become one of the highest priorities in modern surgical practice.[7] Especially, in the field of Congenital Heart Surgery (CHS), where adverse outcomes can be frequent due to the wide spectrum of congenital cardiac conditions and severity of pathologies, as well as high proportion of palliations.[8,9] It is obvious that 30 days mortality measure carries important limitation for assessment of residual pulmonary regurgitation or stenosis, arrhythmia and ventricular failure.

According to the Good Clinical Practice guidelines defined and recommended by World Health Organization (WHO), all database contents should be verified by comparison with the original records at the site of their origin-Source Data Verification (SDV).[10,11] Two patients, out of thirty after verification, have been missed. There have been no statistically significant differences between preoperative sinus rhythm and rhythm at discharge. Undoubtedly, modern intensive care technology extends critical patients survival time what does not necessarily mean increase in long-term survival and improvement of the quality of life. All congenital cardiac surgery 30 days mortality was 3.58%[12].

Conclusions.

Surgical treatment for Tetralogy of Fallot in adult patients at the National cardiovascular Centre Jakarta-Indonesia is quite save and beneficial; a well organized long term follow up is needed. The care of adult patients with corrected or uncorrected CHD is growing and become a challenging new field for the cardiologist in Indonesia

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