Long-term outcome after surgery for pulmonary stenosis (a longitudinal study of 22–33 years)

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KEYWORDS

Pulmonary stenosis; Congenital heart defect; Survival; Follow-up study Aims Long-term (>20 years) survival and clinical outcome are only partly documented in patients who underwent surgical repair for isolated pulmonary stenosis. Yet, such data are of critical importance for the future perspectives, medical care, employability, and insurability of these patients.

Methods and results Ninety consecutive patients underwent surgery for pulmonary stenosis between 1968 and 1980 at the Thoraxcenter. A systematic follow-up study was performed in 1990 and again in 2001. Survival after 25 years was 93%. Re-intervention was necessary in 15% of the patients, mainly for pulmonary regurgitation. Right atrial and ventricular dilatation and paradoxical septal motion were associated with the need for reoperation. No major ventricular arrhythmias occurred. Supraventricular arrhythmias occurred, only in patients with severe pulmonary regurgitation and disappeared after reoperation. At last follow-up, 67% of the patients was in NYHA Class I and maximal exercise capacity was 90% of normal. Moderate or severe pulmonary regurgitation was present in 37% of the patients.

Conclusion Although long-term survival and quality of life are good, pulmonary regurgitation is found in a third of the patients 22–33 years after surgical repair for isolated pulmonary stenosis and reoperation for pulmonary regurgitation was necessary in 9%, especially after the transannular patch technique.

Introduction

Surgical treatment was the only option for valvular pulmonary stenosis before the use of balloon angioplasty. 1 The surgical approach evolved from closed valvulotomy, first performed in 1948, to open valvulotomy using inflow occlusion and finally, open valvulotomy with the use of cardiopulmonary bypass. 2 With the latter technique, the extent of surgery can be adapted to the needs of the individual patient and can vary from a simple commissurotomy, to a complete right ventricular outflow tract reconstruction with a transannular patch or homograft. Current surgery is associated with low early mortality and good short to intermediate term results.^{3,4} However, there are few reports of late outcome of patients, operated upon in 1960s and 1970s, patients who are now adults. 5,6 In fact, biased information has been gathered, because many adult patients have been discharged from routine cardiological follow-up and the adult cardiologist in particular sees patients who developed symptoms. Reliable information about the 'natural history' after surgical valvotomy for isolated pulmonary valve stenosis can only be obtained if an unselected surgical cohort is investigated. Such information

is important with regard to the future perspectives, employability and insurability of these patients. The aim of our study is to determine the ultra long-term survival (22–33 years) and clinical outcome (e.g. incidence of reoperation, residual lesions) of a single centre cohort of consecutive patients operated for isolated pulmonary stenosis between 1968 (start of the heart-lung machine in our centre) and 1980.

Methods

Patients

Ninety patients had surgical repair of an isolated pulmonary stenosis at our institution between 1968 and 1980 at young age (≤15 years). Pre-operative clinical findings, including cardiac catheterization, and details of the surgical procedure were collected from the hospital records of all 90 patients. A first follow-up study was performed in 1990: at this time, 3 patients had died, 51 patients were invited to participate in the study. By an administrative mistake, the other 36 patients were not invited. From the 51 invited patients, 45 (88%) participated in the first follow-up study in 1990. The 45 patients of the 1990 study were invited for a second follow-up study in 2001 of whom 38 participated (*Figure 1*). The cardiac examination in both 1990 and 2001 included medical history, physical examination, standard 12-lead electrocardiography (ECG) (Holter), 24 h ambulatory ECG, echocardiography, and bicycle ergometry.

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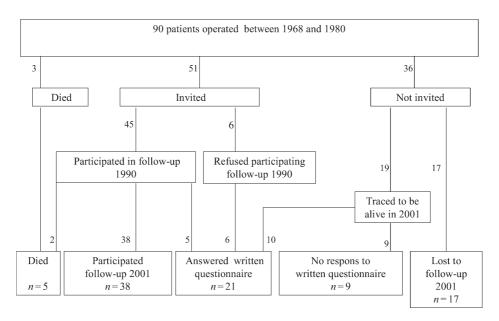


Figure 1 Flowchart of the 90 patients with congenital pulmonary stenosis operated at young age between 1968 and 1980.

The Medical Ethical Committee approved the study. All patients gave their written consent.

In 2001, an attempt was made to locate and contact all 87 patients. Two additional patients had died, 17 were lost and the remaining 68 patients were alive. In addition to the 38 patients who participated in the follow-up study, 21 of the remaining 30 patients returned a written questionnaire with information on morbidity.

Electrocardiography

Standard 12-lead electrocardiograms were analysed for cardiac rhythm, the height and duration of the P-wave (measured in lead II), and the PR interval. A first-degree atrioventricular block was defined by a PR interval >200 ms. Furthermore, the median frontal plane P-wave axis and QRS axis were determined, as was the longest QRS duration. A QRS duration >120 ms was classified as a complete bundle branch block: a positive QRS complex in lead V1 was categorized as right-bundle branch block and a negative QRS complex as left-bundle branch block. A single observer, who was blinded for the echocardiographic results, made all ECG measurements (JR).

Holter monitoring

Sinus node dysfunction was assessed during 24 h Holter monitoring using the modified Kugler criteria: nodal escape rhythm, sinus arrest >3 s or severe sinus bradycardia (<30 b.p.m at night or <40 b.p.m during daytime). Ventricular tachycardia was defined as three or more consecutive ventricular beats with a heart rate of >100 b.p.m.

Echocardiographic measurements and definitions

Pulmonary regurgitation was assessed by colour Doppler and was classified as (1) absent or light, (2) moderate, or (3) severe. Severe regurgitation was defined as a broad regurgitant jet in the right ventricle (RV), with diastolic retrograde flow in the pulmonary artery. Moderate regurgitation was defined as a moderate to broad regurgitation jet, without diastolic retrograde flow in the pulmonary artery. In mild regurgitation, the regurgitant jet was narrow. The RV was defined as severely enlarged if the RV was larger than the left ventricle (LV) in the apical four-chamber view. Moderate enlargement was defined as a larger than normal RV, but maximally

as large as the LV. No differentiation was made between no or mild dilatation. The RV outflow tract was measured by M-mode in the parasternal long axis in end-diastole. The end-diastolic RV inlet diameter was measured in the apical four-chamber view, just above the tips of the tricuspid valve, from the interventricular septum to the lateral wall. The fractional shortening was not calculated and analysed in patients with paradoxical septal movement. Two persons (FM and SS) measured independent from each other, all echo parameters from both the 1990 and the 2001 studies. For echocardiographic measurements of the RV, every patient served as his own control; the measurements of the 1990 study were compared with those of 2001.

Bicycle ergometry

Maximal exercise capacity was assessed by bicycle ergometry with stepwise increments of 20 W/min until exhaustion. Exercise capacity was compared with that in normal individuals corrected for age, sex, and body height.⁸ The results of the 1990 study were compared with the 2001 study.

Data analysis

Continuous data are presented as mean \pm one standard deviation, except patient age and follow-up duration, which are presented as median value and (interquartile) range. Dichotomous variables are presented as counts and percentages. We applied unpaired Student's t-tests (continuous variables) and χ^2 test or Fisher's exact tests (dichotomous variables) to evaluate differences in clinical and echocardiographical characteristics between independent subgroups of patients. Paired Student's t-tests were applied to study changes in continuous variables between the 1990 and the 2001 follow-up visits, whereas the McNemar test of symmetry was applied to study changes in dichotomous variables. The method of Kaplan–Meier was used to estimate cumulative survival during 30-year follow-up. We (graphically) present estimated long-term survival together with the corresponding 95% confidence interval.

This is, basically, a descriptive study, and the results should be interpreted as such. However, despite the fact that we did not have one well-defined research question, multiple significance tests were performed to substantiate specific, clinically relevant statements. As a result, the overall Type I error might be substantial. To provide insight into the level of significance, we present

J.W. Roos-Hesselink *et al.*

actual P-values for each test. Still, we considered a P-value < 0.05 statistically significant. All significance tests were two-sided.

Results

Patients

Median age at operation was 5.0 years (range 0–14). The mean pressure difference between RV and pulmonary artery before surgery was 95 \pm 39 mmHg. The surgical technique is summarized in *Table 1*. The transpulmonary approach was used in most of the patients and a transannular patch was inserted in 13%.

Survival

Peri-operative mortality occurred in two patients. One patient died 10 months after surgical repair as the result of severe brain damage due to an aortic dissection at surgery. Two patients died, respectively, 24 and 25 years after surgery; one patient died during a car accident and the other patient died during his sleep, no autopsy was performed. Late (post-discharge) survival after PS repair was 96% at 25 years (*Figure* 2).

Follow-up

After a median follow-up of 16 years (range 11–22), 45 patients participated in the first follow-up study. Of these, 2 patients died and 38 patients participated in the second follow-up study in 2001 with a median follow-up of 27 years (range 22–33) after surgery and with a median age at the time of study of 32 years (range 22–44). Twenty patients were male (52%) and eighteen female (48%). Twenty-one additional patients returned the written questionnaire and they are all in good clinical health, have not been admitted to the hospital for re-intervention or pacemaker implantation and only one patient have experienced a tachyarrhythmia treated with medication. The baseline characteristics of the patients who did and did not participate in the follow-up study of 2001 are described in *Table 2*.

Re-interventions

From the original cohort of 90 operated patients, we have information on re-interventions in 64 patients. In 10 patients (15%), re-interventions were performed: two patients underwent balloon valvuloplasty because of a substantial

residual pulmonary stenosis, respectively, 16 and 18 years after surgery; eight patients needed a reoperation: two early, respectively, 2 and 3 years after surgery because of residual right ventricular outflow tract obstruction and in six patients a pulmonary homograft was implanted because of severe pulmonary regurgitation late (median 20 years, range 16–24) after the initial operation. Only symptomatic patients were referred for pulmonary valve replacement. In five of these six patients, the surgical technique of the primary operation included a transannular patch, whereas in one, a simple valvotomy was performed. In two of the six patients, in addition, a tricuspid valve plasty was performed and in one an atrial septal defect was closed.

In a separate analysis, we tried to investigate what echocardiographic data would predict a reoperation. Therefore, we compared the echo data of the 1990 study for the patients who needed a reoperation in the next decade, with the echo data of the patients without need for reoperation. The transpulmonary gradient in the six patients who needed reoperation was not different from the patients without reoperation. However, right atrial and right ventricular dilatation and paradoxical septal motion were substantially more frequent in the patients who needed reoperation between 1990 and 2001 (*Table 3*).

Pacemaker

Pacemaker implantation was performed in two patients because of sick sinus syndrome, respectively, 16 and 18 years after surgery.

Clinical evaluation

In 1990, 77% of the patients was in NYHA Class I and 23% in Class II. In 2001, 67% was in NYHA Class I, 30% in Class II, and 3% in Class III ($P\!=\!0.4$). The outcome of patients own health assessment, compared with that of the normal population, is shown in Table 4.9 One patient was taking cardiac medication: oral anticoagulation and a betablocker. Half of the patients had regular follow-up at a cardiology outpatient clinic. Three patients with severe pulmonary regurgitation needed (repeated) electrical cardioversion for atrial fibrillation or flutter. None of these three had a recurrence of arrhythmia after pulmonary valve replacement. No patient showed clinical signs of heart failure. No endocarditis occurred. Of the 18 female patients, nine had successful pregnancies. None of the offsprings had cardiac defects.

Surgical technique	1968-1974	1975-1980	Total period	Median age at operation in years (interquartile range
Number of patients	49	41	90	5.5 (2-8)
Inflow occlusion Transpulmonary approach	33	14	47	7 (4-10)
Cardiopulmonary bypass				
Transpulmonary approach	4	9	13	6 (3-9)
Transventricular approach	10	7	17	5 (3-8)
Transannular patch	1	11	12	1 (0-2)
Homograft RV-PA	1	0	1	10 (–)

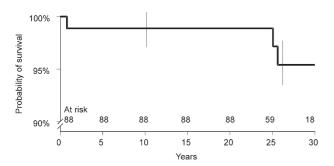


Figure 2 Post-discharge survival after PS surgery. Kaplan-Mayer survival curve of the 88 patients who were discharged after surgery for pulmonary stenosis in our centre between 1968 and 1980.

Table 2 Baseline characteristics of patients with vs. without follow-up in 2001

	With follow-up	Without follow-up
Number of patients	38	52
Age at operation in years (interquartile range)	5.0 (2-7)	6.1 (3-9)
Surgical technique		
Inflow occlusion		
Transpulm	10	37
Cardiopulm bypass		
Transpulm	7	6
Transventric	12	5
Transann patch	8	4
Homograft RV-PA	1	0
Cardioplegia	3	3
RV-pressure pre-operative (mmHg)	125 ±43	103 ± 41
RV-pressure end procedure (mmHg)	54 ± 37	55 ± 37

Electrocardiography

The QRS duration increased from 96 \pm 16 to 108 \pm 22 ms in 11 years time (Table 5). Right-bundle branch block was present in 22%. We found no difference between transpulmonary and transventricular approaches (with right ventricular incision) regarding the incidence of a right-bundle branch block during follow-up. The increase in QRS width was 25 \pm 20 ms for the patients who underwent a reoperation and 11 \pm 18 ms for the patients without reoperation between 1990 and 2001. After successful surgery, no patient showed decrease of the width of the QRS complex. The QRS complex in 2001 was wider for the patients with right ventricular dilatation on echocardiography: 120 \pm 22 ms vs. patients without dilatation: 97 \pm 18 ms. In addition, the increase in QRS width was more: 16 \pm 18 ms for the patients with dilatation vs. $7 \pm 14 \,\mathrm{ms}$ for the patients without dilatation.

Twenty-four hour ambulatory monitoring

None of the patients had atrial flutter or fibrillation in 1990 or 2001 on 24 h ambulatory monitoring. Signs of sinus node disease were found at last follow-up in one patient. No ventricular pauses longer than 3 s occurred. Ventricular

Table 3 Comparison of echocardiographic data from the followup study in 1990 of patients without and with need for reoperation in the next decade (1990–2001)

	Without reoperation	With reoperation	P-value
Number of patients	39	6	0.7
$V_{\rm max}$ pulm valve (m/s)	1.6 ± 0.3	1.7 ± 0.3	0.7
RA dilatation (%)	15	100	< 0.0001
RV dilatation (%)	10	67%	0.02
RVOT (mm)	27 ± 6.5	35 ± 7.1	0.1
RV inlet (mm)	42 ± 8.4	45 ± 8.6	0.5
Paradox septal motion (%)	5	83	<0.0001
Severe TI (%)	26	83	0.01
Severe PI (%)	38	83	0.02

 $V_{\rm max}$ pulm valve, maximal velocity measured with continuous wave Doppler over the pulmonary valve; RA, right atrium; RVOT, right ventricular outflow tract; TI, tricuspid regurgitation; PI, pulmonary regurgitation.

Table 4 Personal health assessment of patients 22-33 years after surgery for pulmonary stenosis compared with the normal Dutch population <35 years

	Patients	Normal Dutch population patients
Numbers	38	1510
Excellent (%)	22	40
Good (%)	70	50
Fair (%)	11	9
Not good (%)	_	1
Bad (%)	-	_

The personal health assessment of 38 patients was compared with 1510 controls of the normal Dutch population, matched for age and sex. There was a significant difference in the health assessment distribution between the patients and the control population (P < 0.05).

tachycardia of more than 10 complexes was not found, whereas 2% showed ventricular tachycardia of 3-10 complexes.

Echocardiography

Pulmonary regurgitation (moderate or severe regurgitation) was present at last follow-up in 37% of the patients (Table 6). Over the last 11 years, two patients showed progression from moderate to severe pulmonary regurgitation. We found no correlation between the presence of pulmonary regurgitation and the age at surgery or right ventricular incision during surgery. However, we did find a correlation with the use of a transannular patch. One patient showed a mild stenosis on the left pulmonary artery (gradient 19 mmHg). The maximal flow velocity over the pulmonary valve did not change significantly in 10 years time $(1.6 \pm 0.3 \,\mathrm{m/s} \, \text{in} \, 1990 \, \text{to} \, 1.7 \pm 0.3 \,\mathrm{m/s} \, \text{in} \, 2001)$. In five patients, the right ventricular dimension increased between 1990 and 2001, in two of these also an increase in pulmonary regurgitation was found, in the other three, pulmonary regurgitation was moderate and remained unchanged. Only two of the six patients who underwent pulmonary homograft implantation showed regression of

J.W. Roos-Hesselink *et al.*

Table 5 Standard 12-lead ECG 22-33 years after surgery for PS

	1990	2001	P-value
Number of patients	45	38	
Rhythm (%)			
Sinus	39 (87)	35 (92)	0.3
Nodal	3 (7)	0	
Atrial	3 (7)	1 (3)	
Pacemaker	0	2 (5)	
Aflutter	_	_	
PR interval (ms)	150 \pm 21	150 \pm 24	0.15
QRS duration (ms)	96 ± 16	108 \pm 22	< 0.0001
RBBB (%)	9	22	0.007
QTc segment (ms)	387 ± 23	392 ± 25	0.6
QRS axis (°)	80 ± 20	63 ± 17	0.08
P-wave duration (ms)	85 ± 11	90 ± 13	0.01
P-wave height (cm)	0.30 ± 0.06	0.20 ± 0.09	0.9
P-wave axis (°)	60 ± 18	56 ± 24	0.1
LVH (%)	0	0	1.0

Comparison of the standard 12-lead electrocardiogram in patients 16 and 27 years after surgery for pulmonary stenosis. Aflutter, atrial flutter; RBBB, right-bundle branch block; LVH, left ventricular hypertrophy.

right ventricular dilatation. Paradoxical septal motion was found in eight patients at last follow-up; 7 of these 8 had severe pulmonary regurgitation. Half of the patients (53%) showed moderate to severe tricuspid regurgitation at last follow-up.

Maximal exercise

The mean exercise capacity was $101\pm21\%$ in 1990 and $90\pm14\%$ of the predicted values in 2001. Almost all patients showed some decrease in exercise capacity between 1990 and 2001. The mean maximal heart rate was in both studies $91\pm10\%$ of expected. No arrhythmias occurred. We found no relation between exercise capacity and velocity over the pulmonary valve before surgery, duration of aortic cross clamp time, presence or absence of a right-bundle branch block on the ECG, or young age (<1 year) at operation. However, the patients with right ventricular dilatation and severe pulmonary regurgitation significantly showed lower exercise capacity (P=0.04). In two of the six patients who underwent reoperation, because of severe pulmonary regurgitation, the exercise capacity improved but in the other four it decreased.

Discussion

This series comprises a cohort of consecutive patients operated at young age at a single institution with longitudinal follow-up of 22–33 years. This study shows low mortality after operation for pulmonary stenosis with residual pulmonary regurgitation being the most important sequel.

Patient selection

In 1990, three patients had died and for administrative reasons, only 51 out of 87 patients were invited for the study of whom 45 (88%) participated. To describe the longitudinal outcome and to have comparable data over time, we have chosen to invite these 45 patients for the complete

Table 6 Echocardiographic results, 22–33 years after surgery for PS

	1990	2001	P-value
Number of patients	45	38	
LA dimension (mm)	32 ± 6	37 ± 5	0.4
LVED dimension (mm)	44 ± 4	46 ± 5	0.7
RA dilatation (%)	24 (53)	24 (63)	0.9
RV dilatation (%)	23 (51)	20 (53)	0.8
RVOT (mm)	28 ± 6	28 ± 6	0.3
RV inlet (mm)	43 ± 8	42 ± 8	0.5
LVH (%)	0	0	1.0
RVH (%)	5 (11)	7 (18)	
Median FractShort LV (%)	35 ± 2	33 ± 2	0.6
Median FractShort RV (%)	20 ± 1	18 ± 1	0.3
Valve insuff (%) PI			
Absent or mild	24 (54)	24 (63)	0.1
Moderate	7 (16)	6 (16)	
Severe	14 (31)	8 (21)	
TI			
Absent or mild	25 (55)	18 (47)	0.2
Moderate	17 (38)	19 (50)	
Severe	3 (7)	1 (3)	
$V_{max}TI$	$2.4\pm~0.3$	2.6 ± 0.3	0.4
V _{max} PS (m/s)	1.6 ± 0.3		0.8

LA, left atrium; LVED, left ventricular end-diastolic; RVH, right ventricular hypertrophy; FractShort, fractional shortening; valve insuff, valve insufficiency; $V_{\rm max}$ TI, maximal velocity measured with continuous wave Doppler of the tricuspid regurgitation; $V_{\rm max}$ PS, maximal velocity measured with continuous wave Doppler over the pulmonary valve.

protocol in 2001. Information on survival, however, was obtained in 73 patients and information on reoperation and hospital admission were gained on 64 patients (21 written questionnaire). There were some differences between the groups with and without complete study protocol. The uninvestigated group tended to have their surgery in an earlier period and had less often the operation with the transannular patch technique. This may have influenced our results with an overestimation of the percentage of reoperations.

Mortality

Late mortality is low in our study. Others report similar excellent long-term survival.^{3,10} Less favourable outcome is found only in patients operated at older age.³ An explanation for this may be the longer period of right ventricular pressure overload with subsequent right ventricular failure.

Residual lesions and re-interventions

Residual pulmonary stenosis was found in two patients needing a re-intervention. In all other patients, only mild stenosis was found at follow-up. Obviously, this was achieved at the cost of pulmonary insufficiency. In 15% of the patients, a re-intervention was necessary. Hayes et al.⁴ found a low (4%) need for reoperation during their study, but their study period was 10 years only. In our study, also only 3% of the patients needed a re-intervention in the first 10 years after surgery and this was for residual pulmonary stenosis. All the other re-interventions were

performed 15 years or longer after the first operation and mainly because of severe pulmonary regurgitation. The need for reoperation was not correlated with the preoperative severity of stenosis, but rather with the surgical technique used and the amount of pulmonary regurgitation, which occurred after surgery. This is similar to the clinical course in patients with tetralogy of Fallot in whom pulmonary regurgitation leads to clinical problems only after a long period of time. Only now are we beginning to realize that the long-term consequences of surgery for pulmonary stenosis are pulmonary regurgitation. The long-standing volume overload of pulmonary regurgitation, leading to right ventricular dysfunction over time, leads to complaints 15-30 years after the initial surgery. At first, compensatory mechanisms based on right ventricular dilatation and paradoxical septal movement develop and only after a longer period, high end-diastolic pressures, secondary tricuspid regurgitation and right-sided heart failure appear. Also other factors such as aging with cell destruction and apoptosis may be responsible for this late right-sided heart failure in ventricles which could cope for so many years. By analysing the echocardiograhic results of 1990, we found that the patients with need for reoperations during the following decade had not only significantly more severe pulmonary regurgitation, but also more often right atrial and right ventricular dilatation, paradoxical septal motion, and severe tricuspid regurgitation. Pulmonary regurgitation occurred especially in the patients who underwent valvuloplasty with use of a transannular patch. The transannular patch technique is a well-recognized risk factor for the development of pulmonary regurgitation in Fallot patients and now proofs to be a risk factor in isolated pulmonary valve stenosis patients as well. 11 This implies that patients after surgical repair of pulmonary stenosis with a transannular patch should be followed carefully, because they are at substantial risk of severe pulmonary regurgitation needing a second (and maybe third) operation. In the future, we should advice our surgeons to use a long small patch to prevent the development of severe pulmonary regurgitation. Information on equally long-term outcome after balloon valvuloplasty is not yet available, but studies with shorter follow-up intervals show good results and this technique is less invasive, less expensive and requires a shorter hospital stay. 12,13 Therefore, at the moment, balloon valvuloplasty is the treatment of choice for patients with isolated pulmonary stenosis. However, long-term follow-up on this treatment modality is warranted and our surgical data should provide a standard for comparing the long-term outcome with that of percutaneous balloon valvuloplasty. In the patients who underwent a reoperation with pulmonary valve replacement because of residual pulmonary insufficiency, right ventricular dimensions diminished in only 2 and remained unchanged in four of the six patients. Maybe, this should be an argument in favour of earlier valve replacement to prevent irreversible right ventricular dilatation. Pulmonary valve replacement late after right ventricular outflow tract obstruction can be performed with low risk and provides clinical improvement in symptomatic patients. However, pulmonary homografts have a limited lifespan. 14,15 The same discussion of when to operate residual pulmonary regurgitation is currently being held for Fallot patients and is still not answered with full satisfaction.

Arrhythmias

Clinical significant arrhythmias occurred only in patients with severe pulmonary regurgitation need of reoperation and, very importantly, they all disappeared after surgery. As in Fallot patients, the supraventricular arrhythmia seems to be the result of elevated right ventricular filling pressures with subsequent right atrial overload. The prevalence of supraventricular arrhythmia may be a marker for the timing of reoperation.

No ventricular arrhythmias occurred and although some comparison with Fallot patients has been made, the duration of the QRS complex is definitely much shorter after surgery for pulmonary stenosis than after tetralogy of Fallot. This supports the theory that the ventricular septal defect (and surgical closure of it) contributes to the widening of the QRS complex in Fallot patients.

Clinical condition

At last follow-up NYHA Class, subjective well-being and exercise performance was lower than expected as is described by others. 17,18 We suggest that pulmonary regurgitation may account for the impairment in clinical condition. 19 Indeed exercise capacity was related to dilation of the RV with severe pulmonary regurgitation. Serial evaluation with stress testing seems a valuable tool to define the moment of re-intervention in patients with pulmonary regurgitation after surgical pulmonary valve repair. In Fallot patients, right ventricular function seems to improve after late pulmonary valve replacement for residual regurgitation. 20,21 In the six patients, in our study, who underwent late reoperation for pulmonary regurgitation, we observed an increase in QRS duration, instead of decrease, and also exercise capacity increased in only two of the six patients, suggesting that probably four of these six patients were reoperated to late. Earlier re-intervention may be warranted in patients with severe pulmonary regurgitation and right ventricular dilatation.

Conclusion

Different surgical management techniques for pulmonary stenosis all lead to a good and long-lasting relief of elevated right ventricular pressure, but this is achieved at the cost of pulmonary regurgitation in a third of the patients. Patients operated upon with a transannular patch are at increased risk for reoperation. Especially right atrial and ventricular dilatation and paradoxical septal motion are predictive for pulmonary regurgitation with need of reoperation. Supraventricular arrhythmias disappeared after reoperation. Pulmonary regurgitation may account for some impairment in clinical condition in these patients.

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J.W. Roos-Hesselink *et al.*

References

- 1. Kan JS, White RI, Mitchell WE, Andersen JH, Gardner TJ. Percutaneous transluminal balloon valvuloplasty for pulmonary valve stenosis. *Circulation* 1984;69:554–560.
- Brock R. The surgical treatment of pulmonary stenosis. Br Heart J 1961;23:337-356.
- Kopecky SL, Gersh BJ, McgoonMD, Mair DD, Porter CJ, Ilstrup DM, Mcgoon DC, Kirklin JW, Danielson GK. Long-term outcome of patients undergoing surgical repair of isolated pulmonary valve stenosis. *Circulation* 1988; 78:1150–1156.
- Hayes CJ, Gersony WM, Driscoll DJ, Keane JF, Kidd L, O'Fallon M, Pieroni DR, Wolfe RR, Weidman WH. Second natural history study of congenital heart defects. Results of treatment of patients with pulmonary valvular stenosis. Circulation 1993;87(Suppl. 1):1-28-1-37.
- Reid JM, Coleman EN, Stevenson JG, Inall JA, Doig WB. Long-term results of surgical treatment for pulmonary valve stenosis. Arch Dis Child 1976:51:79–81.
- Morris CD, Menashe VD. 25-Year mortality after surgical repair of congenital heart defect in childhood. JAMA 1991;266:3447–3452.
- 7. Kugler J. Sinus node dysfunction. *The Science and Practice of Pediatric Cardiology.* 1st ed. Lippincott Williams & Wilkins; 1990:1751–1785.
- 8. Wasserman K, Hansen JE, Sue DY, Stringer WW, Whipp BJ. In: Philadelphia, ed. *Principles of Exercise Testing and Interpretation*. 2nd ed. Lea and Febiger; 1994.
- Netherlands Central Bureau of Statistics. Continuous Survey Quality of Life. Voorburg/Heerlen: Netherlands Central Bureau of Statistics; 2002.
- McNamara DG, Latson LA. Long-term follow-up of patients with malformations for which definitive surgical repair has been available for 25 years or more. Am J Cardiol 1982;50:560-568.
- Kirklin JK, Kirklin JW, Blackstone EH, Milano A, Pacifico AD. Effect of transannular patching on outcome after repair of tetralogy of Fallot. *Ann Thorac Surg* 1989;48:783-791.
- Witsenburg M, Talsma M, Rohmer J, Hess J. Balloon valvuloplasty for valvular pulmonary stenosis in children over 6 months of age: initial results and long-term follow-up. Eur Heart J 1993;14:1657–1660.

- 13. Peterson C, Schilthuis JJ, Dodge-Khatami A, Hitchcock JF, Meijboom EJ, Bennink GB. Comparative long-term results of surgery versus balloon valvuloplasty for pulmonary valve stenosis in infants and children. *Ann Thorac Surg* 2003;**76**:1078-1082.
- Kanter KR, Budde JM, Parks WJ, Tam VK, Sharma S, Williams WH, Fyfe DA.
 One hundred pulmonary valve replacements in children after relief of right ventricular outflow tract obstruction. *Ann Thorac Surg* 2002; 73:1801–1806.
- Willems TP, Bogers AJ, Cromme-Dijkhuis AH, Steyerberg EW, van Herwerden LA, Hokken RB, Hess J, Bos E. Allograft reconstruction of the right ventricular outflow tract. Eur J Cardiothorac Surg 1996; 10:609–614
- Roos-Hesselink JW, Perlroth MG, McGhie J, Spitaels SEC. Atrial arrhythmias in adults after repair of tetralogy of Fallot. Correlations with clinical, exercise and echocardiographic findings. *Circulation* 1995; 91:2214–2219.
- Driscoll DJ, Wolfe RR, Gersony WM, Hayes CJ, Keane JF, Kidd L, O'Fallon M, Pieroni DR, Weidman WH. Cardiorespiratory response to exercise of patients with aortic stenosis, pulmonary stenosis and ventricular septal defect. Circulation 1993;87(Suppl. I):I-102-I-113.
- Yetman AT, King S, Bornemeier RA, Fasules J. Comparison of exercise performance in patients after pulmonary valvulotomy for pulmonary stenosis and tetralogy of Fallot. Am J Cardiol 2002;90:1412-1414.
- Rosenthal M, Bush A. The effects of surgically treated pulmonary stenosis on lung growth and cardiopulmonary function in children during rest and exercise. Eur Respir J 1999;13:590-596.
- Eyskens B, Reybrouck T, Bogaert J, Dymarkowsky S, Daenen W, Dumoulin M, Gewillig M. Homograft insertion for pulmonary regurgitation after repair of tetralogy of fallot improves cardiorespiratory exercise performance. Am J Cardiol 2000;85:221–225.
- 21. Vliegen HW, vanStraten A, deRoos A, Roest AA, Schoof PH, Zwinderman AH, Ottenkamp J, vander Wall EE, Hazekamp MG. Magnetic resonance imaging to assess the hemodynamic effects of pulmonary valve replacements in adults late after repair of tetralogy of Fallot. Circulation 2002;106:1703-1707.