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Long term follow-up after the Ross procedure (RCD code: IV-5A.O)

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Abstract

Aims: The aim of this study was to analyze the incidence of long-term complications observed during follow-up of patients who had undergone the Ross procedure in childhood. **Methods:** The study engaged a cohort of 9 patients, all of whom were between 19 to 32 years old. Patients had been in the care of the Centre for Rare Cardiovascular Diseases since the age of 18. Clinical and echocardiographic data were collected from the follow-up period. **Results:** 78% of patients had preserved global systolic function of the left ventricle, and 56% had dilatation of the ascending aorta. Due to the aneurysm of the ascending aorta one patient required the Bentall de Bono procedure. Another patient underwent a reoperation because of endocarditis of the pulmonary homograft with severe aortic and pulmonary regurgitation. 1/3 of the studied patients were being considered for a reoperation. 23% of patients developed severe pulmonary valve regurgitation, 33% moderate. 67% of patients developed mild to moderate pulmonary valve stenosis. Most patients were in NYHA class I-II. **Conclusion:** Late complications are frequent in this group of patients, and hence they require surveillance in specialized Centers for Grown-up Congenital Heart Diseases. JRCD 2016; 3 (1): 9–13

Key words: rare disease, aortic valve replacement, pulmonary valve replacement, echocardiography

Background

The first step in the treatment of severe congenital aortic stenosis is transcatheter or surgical valve widening [1]. Aortic valve disease is particularly demanding task for cardiac surgeons. The Ross procedure is considered an excellent alternative for children and selected adult patients requiring elective aortic valve replacement. The Ross procedure is a type of specialized aortic valve surgery, where a patient's diseased aortic valve is replaced with his or her own pulmonary valve (autograft). Dr Donald Ross performed this procedure on humans for the first time in 1967. Earlier, in 1960, Lower et al. described the feasibility of replacing the aortic valve of dogs with the native pulmonary valve. The Ross-Konno procedure is the technique for treatment of complex multi-level left ventricular outflow tract obstruction with severe annular hypoplasia and a dysplastic aortic valve.

Among children and young adults, as well as older yet particularly active patients, the Ross procedure offers several advantages over traditional aortic valve replacement with manufactured prostheses. Longevity of the pulmonary autograft in the aortic position is superior to bioprostheses, such as porcine valves, which tend to degenerate after only a few years in patients under 35 years of age. Pulmonary autografts can grow with the patient and present lower risk of infection, do not cause haemolysis, and offer excellent haemodynamic profile that allows for the reversal of left ventricular remodeling [2-5]. Furthermore, anticoagulation is not required as in mechanical valves. Therefore, individuals are able to lead an active life without the risks associated with anticoagulation therapy. This is especially important for women in childbearing age, who require aortic valve replacement, because anticoagulation is contraindicated in pregnancy.

Many medical centers report very good medium- and long-term outcomes of the Ross operation, predominantly in terms of a high survival rate as compared to other surgical methods [6,7]. Based on data from registry of the Ross procedure the operative mortality before the age of 20 is estimated between 0–11% (median 1.8%) [1]. Quality of life and the physical capacity following the Ross procedure is good, and comparable to healthy individuals [8,9].

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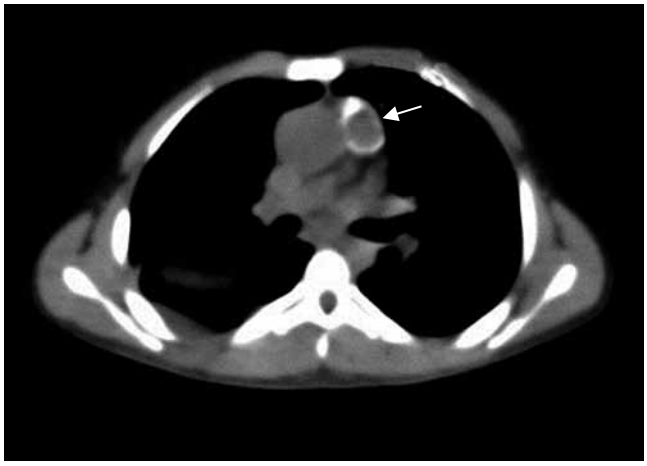

Figure 1. Single-photon emission computed tomography utilizing 99mTc-HMPAO (exametazime) labeled leukocytes. Increased accumulation of marker in the region of pulmonary trunk with visible multiple calcifications – the image seems to correspond to the presence of inflammatory process in this area (arrow)

The autograft valve failure is a possible complication during follow-up. Following the Ross operation, almost 1/3 of the cohort group had dilation of the autograft in aortic position, as well as the autograft valvular insufficiency (especially in patients with the autograft enlargement) in the long-term follow-up [10]. In this study we analyze the incidence of long-term complications during follow-up among patients who underwent the Ross procedure in childhood and are in the care of the Centre for Rare Cardiovascular Diseases.

Methods

The registry of our Adult Congenital Heart Disease Clinic has 9 patients enrolled, two females and seven males. All subjects underwent the Ross procedure in childhood due to a congenital aortic valve stenosis/regurgitation. Among 1/3 of patients a valvulotomy was performed prior to the Ross procedure.

Table 1. Transthoracic echocardiography – comparison of parameters

Patient	LVEF [%]	LV diastolic diameter [mm]	Ao asc [mm]	AV max gradient [mmHg]	AV mean gradient [mmHg]	AV regurgitation	PV max gradient [mmHg]	PV mean gradient [mmHg]	PV regurgitation
1 (mechanic AV, PV- homograft)	60	58	35	19	13	no	3		no
2	60	45	35	12	8	moderate	33	24	severe
3 (PV-homograft)	55	45	31	18	11	mild	25	15	moderate/ severe
4 (Ao asc prosthesis; mechanic Ao Valle; PV-homograft)	25	69	34	7,5	6,5	constructional	14	8,8	moderate
5 (PV-homograft)	55	52	42	4,5	2	mild	32	19	no
6	50	65	61	22	10	moderate	31	19	mild
7 (AV-neoaortic, PV-homograft)	35	68	48	7,5	5	moderate/severe	48	32	moderate
8 (AV-neoaortic, PV-homograft)	61	52	46	6	3	no	53	30	moderate
9 (AV-neoaortic, PV-homograft)	60	48	46	8	4	moderate	74	48	moderate
mean	51±12,8	56±9,6	42±9,4	12±6,4	7±3,8		35±21,2	24±12,2	
9 patients (control group)	LVEF [%]	LV diastolic diameter [mm]	Ao asc [mm]	AV max gradient [mmHg]	AV mean gradient [mmHg]	AV regurgitation	PV max gradient [mmHg]	PV mean gradient [mmHg]	PV regurgitation
mean	64±3,9	46±5,0	26±3,0	6±1,4	3±1,0	no	4±1,7	2±1,4	no

AV- aortic valve, PV-pulmonary valve, LV- left ventricle, LVEF- left ventricular ejection fraction, Ao asc- ascending aorta, F-female, M-male

Figure 2. Computed tomography. Enlargement of the aortic root (46 mm), ascending aorta (61 mm) and aortic arch (40 mm)

The patients have remained in the care of the Centre for Rare Cardiovascular Diseases since the age of 18. During the follow-up, physical examination and transthoracic echocardiography (TTE) were performed by the same medical personnel approximately every 6 and 12 months, respectively. The echocardiographic studies were conducted according to the guidelines of American Society of Echocardiography.

Results

During the follow-up, 78% of the patients had preserved global systolic function of the left ventricle. Among the cohort 56% of the subjects had a dilatation of the ascending aorta with a diameter in the latest follow-up ranging up to 61 mm. Due to ascending aortic aneurysm one patient required reoperation and underwent the Bentall de Bono procedure. Another patient underwent a reoperation because of endocarditis of the pulmonary homograft, with severe aortic and pulmonary regurgitation (Figure 1). One patient with enlargement of the aortic root (46 mm), the ascending aorta (61 mm) and the aortic arch (40 mm) and the right ventricular outflow tract obstruction (RVOTO) was qualified for the Bentall de Bono and a pulmonary valve replacement, however he did not agree to the treatment (Figure 2). Among our patients, 1/3 of the cohort were in the phase of qualification for reoperation. 23% of the patients developed severe pulmonary regurgitation, 33% moderate and 67% developed mild to moderate pulmonary valve stenosis. In 30% of the cohort with preserved left ventricular ejection fraction myocardial perfusion imaging (MPI) demonstrated exercise-induced perfusion defects in the anterior wall segments. No coronary disease was detected by computed tomography angiography in any patients. All the surviving patient remained in New York Heart Association (NYHA) functional class I–II (Table 1,2).

Discussion

The Ross procedure fulfills the initial assumptions about the long-term patients' survival and no necessity for a use of anticoagulation [11-13]. The pulmonary autograft is ideal when it comes to haemodynamics, it does not require anticoagulation and has low thromboembolic risk. Consequently, a concern regarding autograft and pulmonary homograft longevity has appeared. Progressive autograft dysfunction, especially after more than eight years following surgery, indicates the necessity of systematic echocardiographic monitoring in this population [10].

In our study, all patients were clinically asymptomatic and able to perform normal physical activities. It suggested normal myocardial perfusion, without clinical signs of ischemia. Since the final step of the Ross-Konno procedure involves the right ventricular outflow tract reconstruction with the pulmonary autograft, there is a risk of allograft kinking, coronary artery compression or anastomotic stricture due to the location of pulmonary bifurcation posterior to the neoaorta. Similarly, because of the proximity of the septal perforating branches of the left anterior descending artery (LAD) to the suture line, there are chances of septal perfusion getting compromised. Therefore, MPI can be used in the follow-up period to assess an early diastolic dysfunction and LAD territory ischemia. There is, however, no data on the use of MPI especially in this subset of patients in the available literature [14].

56% of our patients had dilatation of the ascending aorta. Pulmonary autograft dilatation is common after the Ross procedure

Table 2. Characteristics of the study group

Patient	Sex [F/M]	Age [years]	Age of the Ross procedure (years)	Reoperation	Type of reoperation	Current complication	NYHA class
1 (mechanical AV, PV- homograft)	M	19	11	Yes; endocarditis of the pulmonary homograft, severe aortic and pulmonary regurgitation	aortic valve replacement (mechanical aortic valve and pulmonary homograft)	history of pulmonary homograft endocarditis, history of sepsis	II
2	M	32	13	No		history of pulmonary homograft endocarditis, RVOTO and severe pulmonary homograft regurgitation	I
3 (PV-homograft)	F	22	9	Yes; pulmonary autograft dysfunction (at the age of 17)	pulmonary homograft replacement	increased RVOT pressure gradient, moderate/severe pulmonary regurgitation	II
4 (Ao asc prosthesis; mechanical AV; PV-homograft)	M	28	12	Yes; aneurysm of the ascending aorta	Bentall de Bono		II
5 (PV-homograft)	M	26	10	No		prosthetic extension of the aortic root	I
6	M	21	3	No; enlargement of the aortic root (46mm), ascending aorta (61mm) and aortic arch (40mm); RVOTO; patient rejected surgery		enlargement of the aortic root (46mm), ascending aorta (61mm) andaortic arch (40mm); RVOTO	I/II
7 (AV-neoaortic, PV-homograft)	M	22	10	the the patient is considered for reoperation		enlargement of the aortic root (49mm), ascending aorta (48mm); moderate/severe neoaortic regurgitation and moderate pulmonary regurgitation; RVOTO	I/II
8 (AV-neoaortic, PV-homograft)	M	27	14	patient is considered for reoperation		enlargement of the ascending aorta (46mm); RVOTO; moderate pulmonary regurgitation	I
9 (AV-neoaortic, PV-homograft)	F	25	9	patient is considered for reoperation		enlargement of the ascending aorta (46mm); RVOTO; moderate aortic and pulmonary regurgitation	II

AV – aortic valve, PV – pulmonary valve, RVOTO – right ventricular outflow tract obstruction, M – male, F – female, NYHA – New York Heart Association functional class

among late survivors and it is the reason for reoperations in this group of patients. The dilatation progresses over time and is often accompanied by enlargement of the native aorta and a secondary valve regurgitation [15]. It results from the lack of leaflets' coaptation caused by changes in the neoaortic root's geometry [11]. Patient age, valve disease pathogenesis, and preoperative aortic regurgitation and dilatation are the most commonly reported patient-related determinants of the durability of an autograft valve. Younger patient age was previously implicated to be associated with increased autograft dilatation but not with late autograft dysfunc-

tion [16,17]. A report by Ruzmetov M et al. showed that a dilatation of the pulmonary autograft was the most common indication for autograft reoperation, with a median interval of 8 years after the original operation [18].

In the Ross procedure a biological valve needed to reconstruct the right ventricular outflow tract, can also degenerate with time. Most surgeons use pulmonary valve homografts for this purpose. Age is the most important determinant of the pulmonary homograft's failure [19-21].

Another important complication of the Ross procedure is infective endocarditis [11]. In our cohort, 23% of patients had endocarditis. Infective endocarditis in patients with a congenital heart disease is associated with almost 10% mortality. Among GUCH patients the proper oral hygiene and antibiotic prophylaxis is important in groups of the highest risk of endocardits. In case of unsuccessful pharmacotherapy, surgery should be performed when serious hemodynamic complications and high risk of septic embolism develops [22,23].

Thromboembolism is an uncommon complication in patients who underwent Ross procedure. It is probably related more to other factors than to the valve itself [19].

Patients after the Ross procedure need a close long-term systematic follow-up, in order to prevent any complications concerning the pulmonary autograft and homograft. These patients require surveillance in centers of reference that specialize in congenital heart diseases.

The Ross procedure provides satisfactory results among children and young adults. Limitations usually appear by the end of the fist postoperative decade, particularly in younger patients [16]. The Ross procedure, when implemented in experienced centers, remains an excellent alternative to conventional aortic valve replacement in certain patients (pediatrics, young adults, women in childbearing age).

Conclusion

Late complications are frequent in this group of patients and therefore surveillance conducted by specialized Centers for Grown-up Congenital Heart Diseases is mandatory.

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