PERCUTANEOUS IMPLANTATION OF A BIOPROSTHESIS IN THE PULMONARY POSITION IN PATIENTS WITH CONGENITAL HEART DEFECTS – EXPERIENCE OF ONE CENTER

Perkutánna implantácia bioprotéza do pulmonálnej pozície u pacientov s vrodenou chybou srdca – skúsenosť jedného centra

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Abstract
Congenital heart diseases (CHD), in which the right ventricle outflow tract (RVOT) was operated in different ways in childhood, will be indicated for reintervention later. Such CHD generate legitimate complications and their the management in most cases belongs to the competence of an adult cardiologist. The prototype of the finding in such an adult patient is severe pulmonary regurgitation, as a natural consequence of the complete correction of Tetralogy of Fallot (TOF) in childhood. There are two options for managing the „common denominator" of different types of RVOT. The first and preferred is a catheterization method – percutaneous pulmonary valve implantation (PPVI), if technically feasible. The second remains the classic cardiac surgery. The PPVI was successfully implemented for the first time in Slovakia in June 2021. By the end of 2024, PPVI had been successfully provided in 20 adult patients and this method has been proved as safe, effective, but especially less invasive alternative to cardiac surgery in candidates. PPVI has thus become an accepted and integral part of solving RVOT dysfunction in Slovakia not only in patients with TOF, but also in less frequent CHD *(Tab. 2, Fig. 15, Ref. 24). Text in* PDF www.lekarsky.herba.sk.
KEY WORDS: tetralogy of Fallot, pulmonary regurgitation, indications for reintervention, percutaneous implantation of pulmonary valve, congenital heart defects in adulthood.
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Abstrat
Vrodené chyby srdca (VCHS), u ktorých bol v detstve rôznym spôsobom riešený výtokový trakt pravej komory (RVOT), časom dospejú do potreby reintervencie. VCHS teda logicky generujú legitímne komplikácie a ich manažment už spadá už vo väčšine prípadov do kompetencie dospelého kardiológa. Prototypom nálezu u dospelého pacienta je závažná pulmonálna regurgitácia, ako prirodzený následok korekcie Fallotovej tetralógie (TOF). Existujú dve možnosti manažmetu „spoločného menovateľa" rôznych typov RVOT. Prvým a preferovaným je katetrizačné riešenie – perkutánna implantácia chlopne do pulmonálnej pozície (PPVI), ak je technicky uskutočniteľná. Druhým zostáva klasická kardiochirurgia. PPVI bola prvýkrát na Slovensku úspešne realizovaná v júni roku 2021. Do konca roku 2024 bolo úspešne odimplantovaných 20 pacientov a táto metóda sa preukázala ako bezpečná, efektívna, najmä menej invazívna a alternatíva chirurgického riešenia. PPVI sa stala aj na Slovensku akceptovanou a neoddeliteľnou súčasťou riešenia dysfunkcie RVOT nielen u pacientov s TOF, ale u menej častých VCHS *(tab. 2, obr. 15, lit. 24). Text v PDF www.lekarsky.herba.sk.*
KĽÚČOVÉ SLOVÁ: Fallotova tetralógia, neskoré komplikácie, indikácie na reintervenciu, perkutánna implantácia pulmonálnej chlopne, vrodené chyby srdca v dospelosti.

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Introduction

CHD is defined as a cardiovascular anomaly present from birth. The incidence at the global level (17.9/1000 births, 19.1/1000 for the male and 16.6/1000 for the female) remains stable in both genders (1). Advances in pediatric cardiology and cardiac surgery have led to a significant improvement in almost 95% survival of children with CHD into adulthood. Thus, the number of adult patients with CHD is currently greater compared to the paediatric population (2). Each CHD carries its own specifics into adulthood, which in most cases eventually need reintervention. Predominantly, these residual findings concern the right heart, but left-sided findings must not be ovelooked either. The most common such

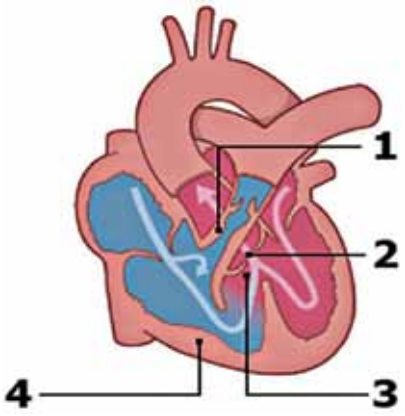
finding is severe pulmonary regurgitation („free PuR“) in patients with TOF. This is a natural consequence of complete correction of the CHD, which results in dilatation of the right ventricle (RV), eventually its dysfunction. If the patient meets the indications for further reintervention, there are two options. According to the latest European Society of Cardiology (ESC) Guidelines for CHD in Adulthood (ACHD) 2020 (3), the first and preferred method is – percutaneous pulmonary valve implantation (PPVI). The second option remains of surgical replacements in majority of patients with a bioprosthesis, and only in exceptional cases with a mechanical prosthesis (3). For many patients, that means reoperation, which is risky in itself. PPVI is safe and effective method, fully established in Slovakia.

Tetralogy of Fallot and other „Fallot-like“ CHD

The anatomy corresponding to the tetralogy of Fallot (TOF) was described by the Danish bishop and physician Stenson (4) in 1673. The clinical syndrome was first described and distinguished from others CHD in 1988 by the French physician Etienne – Louis Arthur Fallot (5). It accounts for about 4-10% of all CHD and represents the most common cyanotic CHD (6). TOF itself is indicated for complete surgical repair in infancy. Within the wide range of CHD, there are different variations of it, which is beyond the scope of this publication.

This complex CHD consists of 4 anatomical alterations (Figure 1).

Figure 1. Tetralogy of Fallot.



1. **pulmonary stenosis (PS):** can be localized subvalvular, valvular, less often supravalvular, at the site of pulmonary arteries and variously combined
2. **ventricular septal defect (VSD):** large, anteriorly malaligned, non-restrictive
3. **overriding aorta over the VSD**
4. **right ventricular hypertrophy (RV):** as a secondary consequence

Complete surgical repair of TOF consists of removal of the RVOT obstruction (RVOTO) by resecting its hypertrophied muscle and extending it, e.g. patch, pulmonary valve excision and VSD closure with a patch. In some cases (significantly narrow RVOT, coronary arteries abnormalities), a conduit of bovine/pork origin or a human homograft can be used as the connection between RV and a. pulmonalis. However, these artificial connections degenerate over time, which leads to the need for further intervention after about 15 years.

„Fallot-like“ CHD

Among others surgically corrected CHD similarly to TOF, so-called.“ Fallot-like „, i.e.: pulmonary atresia, double RV outlet, truncus arteriosus communis, condition after Ross surgery.

Follow up of an adult patient with CHD

Despite the complete surgical repair of the CHD, the defects may not be definitively resolved. So, CHD as a rare disease, require lifelong follow-up in a specialized centre with sufficient experience.

After reaching adulthood, these patients should be transferred to the cardiologist care for adult. According to the recommendations of the ESC for ACHD one specialized center for approx. 5mil. inhabitatnts is sufficient. This model was established by Slovak Ministry of Health 13 years ago (ACHD Centre), which was in 2020 transformed in separate ACHD Department with its own specialized outpatient clinic.

Residual findings in adulthood

The most common residual finding in adulthood in patients with TOF is free PuR, which results in chronic volumetric overload of the RV. This natural consequence of complete repair, needs to be reintervention provided early enough to prevent RV failure.

Pulmonary stenosis and conduit dysfunction usually persist in a minority of patients, it can also affect individual branches of the lungs. If it progresses, RV responds to increased pressure by remodelling in terms of hypertrophy and over time by decrease its systolic function.

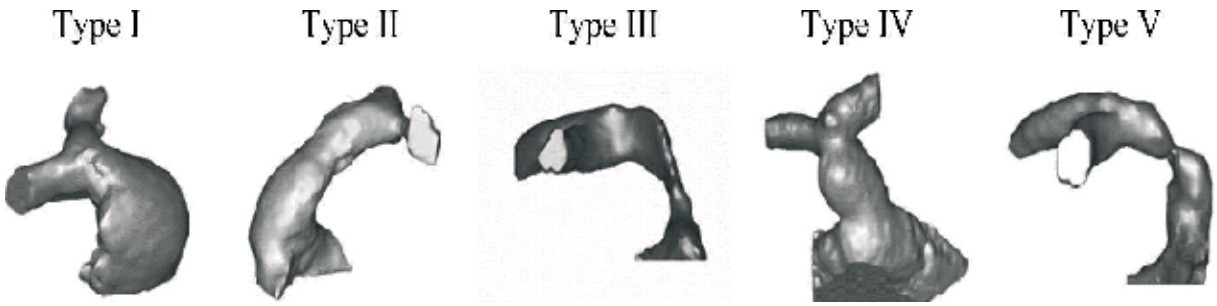
Percutaneous pulmonary valve implantation (PPVI)

The pioneer of this interventional method was Boenhoffer (7), who first to implanted the Melody bioprosthesis in a 12-year-old boy, originally with pulmonary atresia in infancy corrected with conduit in 2000. Since then, this method has rapidly expanded worldwide.

In Slovakia, the PPVI was successfully implemented for the first time in 2021 in cooperation with the ACHD Department and the Department of Functional Diagnostics of Children´s Cardiac Centre of National Institute of Cardiovascular Diseases. 20 patients successfully underwent this procedure by the end of 2024 year.

The indication of PPVI includes several CHD conditions, in which the finding progresses over time and requires invasive reintervention. The decisive site for assessing the anatomical suitability of the PPVI is the RVOT, which represents the variable area of the heart. In Figure 2, several types of it are demonstrated. These were defined on the 3D reconstruction basis of using magnetic resonance imaging and mathematical operations (8).

Figure 2. RVOT types (8).



Explanatory notes:
Type I – RVOT has a pyramidal shape, most common in patients who have received a transannular patch
Type II – RVOT has a constant diameter
Type III – RVOT has the shape of an inverted pyramid
Type IV – RVOT wide in the central part
Type V – RVOT is narrow centrally, but proximal and distal it is wide.

In practice, there are **different types of RVOT**

- **native** - pulmonary valve is completely excised,
- **conduit/homograft** – foreign material tube shaped used as a connection between the RVOT and a.pulmonalis,
- **bioprosthesis** in pulmonary position.

Indications for reintervention in patients with TOF and „Fallot-like" CHD

As part of comprehensive management and regular monitoring, these examinations are usually carried out to tacle with indication for reintervention– echocardiography, ECG Holter, ergometry, examination of NTproBNP levels, CT and MRI, or cardiac catheterization.

According to the 2020 ESC ACHD Guidelines, pulmonary valve replacement (PVR) is recommended for symptomatic patients, a transcatheter procedure should be preferred, if anatomically feasible (Class I, Evidence Level C) (3).

The objective assessment of the patient´s symptoms is still challenging, as they are often young and active in sports with a large functional reserve of the body.

PVR should be considered in **asymptomatic patients** with severe PR or RVOTO when one of the following criteria is present (Class IIa, the level of evidence C) (3):

- the decrease in objective capacity,
- progressive RV dilatation to RV ensdystolic volume indexed ≥ 80 ml/m^2 and/or RV enddiastolic volume indexed ≥ 160 ml/m^2 ,
- tricuspid regurgitation (TR) progresses to at least moderate,
- progressive RV systolic dysfunction,
- residual RVOT obstruction (RVOTO) of > 80 mmHg is present.

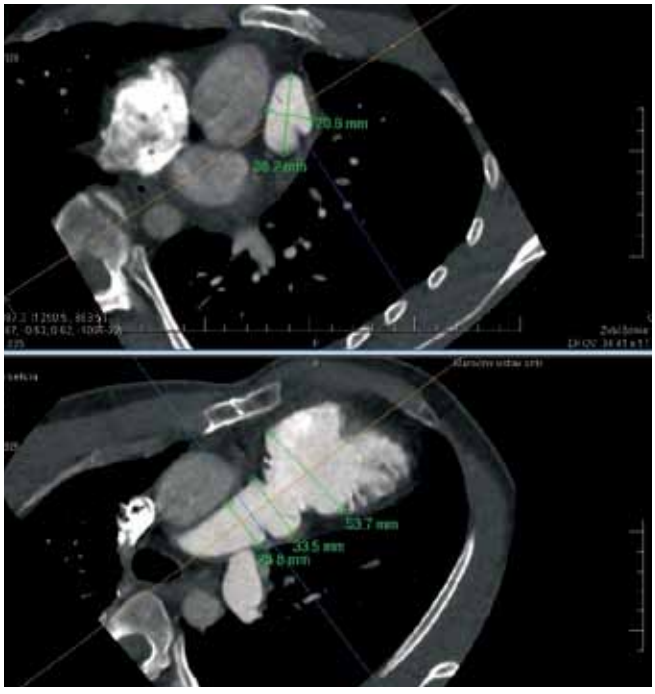
Preparing the patient for PPVI consists of assessing the patient's health status, including comorbidities, not only cardiac issue. The procedure takes place under general anesthesia, the patient undergoes preparation as before the operation.

PPVI procedure

PPVI is done by inserting a catheter system through a large vein (typically the femoral vein). Angiography is used to identify the anatomy of the RVOT and its relation to coronary arteries. Before the valve is implanted the right heart catheterization and measurement of hemodynamic is performed.

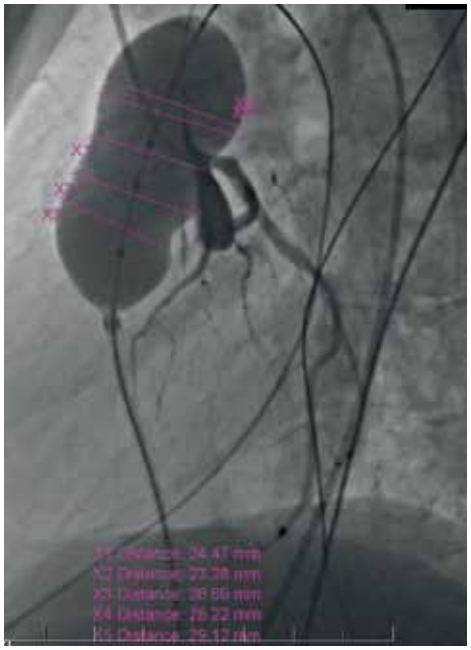
This is followed by imaging and measuring the RVOT in at least two projections with simultaneous comparison and in a CT image (Figure 3) to visualize its minimum size and balloon position.

Figure 3. Measurement of RVOT in at least two positions (Tomocon program, National Institute of Cardiovascular diseases).



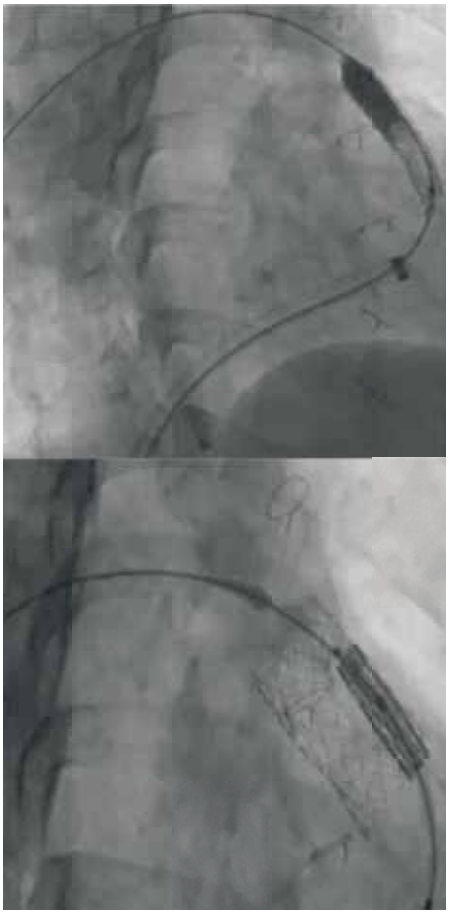
Part of the procedure itself is **the so-called RVOT occlusion test with simultaneous selective coronary angiography.** The purpose of the test is to exclude stent compression of the coronary artery after implantation of the bioprosthesis (Figure 4).

Figure 4. Angiogram shows balloon measurement and coronary artery test (Tomocon program, National Institute of Cardiovascular diseases).



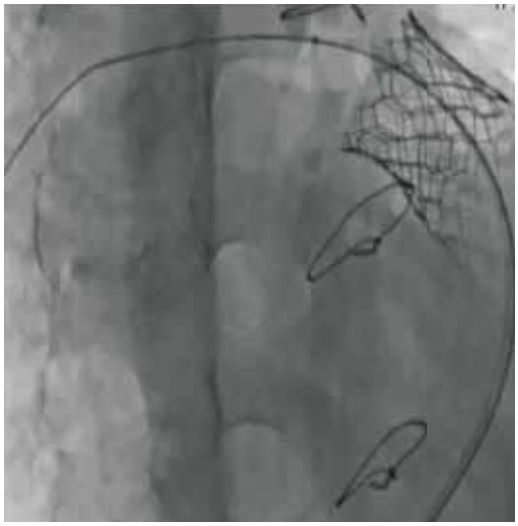
The next step is the preparation of the implantation site**, the so-called „landing zone" = pre-stenting**, followed by the valve implantation itself (Figure 5). The last step is checking the valve function.

Figure 5. Landing zone preparation and pre-stenting (Tomocon program, National Institute of Cardiovascular diseases).



After the procedure, the patient is monitored 24 hours at intensive care unit. If any complication occurs, the total length of hospitalization usually does not exceed 5 days (Figure 6).

Figure 6. Checking the functionality of the valve (Tomocon program, National Institute of Cardiovascular diseases).



In all patients after PPVI, dual antiplatelet therapy (DAPT - acetylsalicylic acid, clopidogrel) for 6 months and then lifelong acetylsalicylic acid is indicated. According the ESC Guidelines for Infective Endocarditis (IE), these patients belong to the highest risk group and therefore the prevention of IE is indicated for life (9).

Types of bioprostheses used in PPVI

The Melody **bioprosthesis** (Medtronic, Minneapolis, MN, USA) was used first. It is of bovine origin, sutured within a platinum-iridium frame dimensions 28mm x18mm, diameter max. 22 mm (Figure 7) (10).

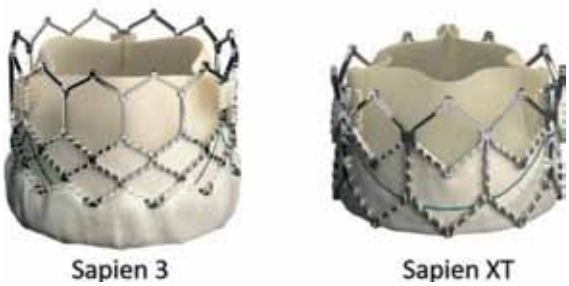
Figure 7. Bioprosthesis Melody (10).

Edwards Sapiens (Edwards Lifesciences, Irvine, CA, USA) is a 3-point bioprosthesis, made of bovine pericardium, implanted using a balloon expandable stent measuring 23 and 26 mm. The Sapient XT is sutured within a cobalt-chromium frame in the dimensions of 20 mm, 23 mm, 26 mm, 29 mm (Figure 8) (11).

The RVOT dimension in patient with CHD is often much wider than the dimensions of the bioprostheses used, and this is the main limiting factor of PPVI.

However, bioprostheses of larger dimensions are gradually being designed and introduced into practice. One of them is the Venus P bioprosthesis.

Figure 8. Bioprosthesis Edwards Sapiens (11).



Venus P (Venus MedTech, Shanghai, China)

It is a self-expandable porcine pericardium valve with diameters ranging from 20 to 32 mm in 2- mm increments., designed for use in RVOTs that have been reconstructed by patching (Figure 9) (12).

Figure 9. Bioprosthesis Venus P (12).

Material and methods

The cohort consists of 20 patients with CHD who met the indication criteria according to the 2020 ESC Guidelines for CHD (3) for further treatment of PV. Part of the patients was indicated by the ACHD Dept. and part by Children's Cardiac Centre of the National Institute of Cardiovascular Diseases.

Majority of patients (n=18) received a 23, 26 or 29 mm Edwards Sapiens bioprosthesis implanted in one session after Andrastent XXL prestening. In one patient, the implantation of the valve for unstable RVOT was performed in two sessions: a stent was implanted in the first procedure and the bioprosthesis itself was implanted after 3 months. Two patients have only recently been implanted with the Venus P bioprosthesis and have not yet undergone an outpatient check-up.

Postprocedurally, patients underwent an outpatient follow-up examination after 3 months.

Selected monitored parameters before and 3 months. after the PPVI were as follows:

- NYHA Functional Classification,
- NTproBnP level examination,
- change in the severity of PuR,
- change in maximum gradient to PV assessed by echocardiography,
- change in indexed enddiastolic and endsystolic volume and RV function assessed by MRI (volumetry).

In all patients after PPVI, dual antiplatelet therapy for 6 months was indicated in accordance with the protocol and recommendations, followed by lifelong acetylsalicylic acid and prevention of IE.

All patients remain in the dispensary of the outpatient clinics of ACHD Dept. and Children´s Cardiac Centre of the National Institute—of Cardiovascular Diseases

Statistical analysis

Due to the small number of patients in the cohort, statistical evaluation was limited, and for this reason descriptive statistics were used. Also, in the individual parameters (MRI, NTproBnP), the results of only patients who managed to undergo it are listed.

The parameters of descriptive statistics are given as absolute numbers, average, minimum and maximum value, individual patients in graphs with initials.

Results

The characteristics of the group of patients by CHD and gender are presented in Table 1. 14 patients from the ACHD Dept. and 6 patients from the Children´s Cardiac Centre underwent the PPVI. The average age was 29 (14 - 70) years. In addition to the complete correction in childhood, minimum 6 patients underwent at least one operation.

Table 1. Characteristics of the patient population by CHD and gender.

CHD	ACHD Dept.		Children´s Cardiac Centre	
	men	women	men	women
TOF	4	3	4	_
PA	2	_	1	_
DORV	0	_	_	_
D-TGA	2	_	_	_
TAC	2	_	_	_
Ross's op.	1	_	1	_

Notes: TOF – Tetralogy of Fallot, PA – pulmonary atresia, DORV – double outlet right ventricle, D – TGA – transposition of large arteries, TAC – truncus arteriosus communis, ACHD Dept. – Department of Adult Congenital Heart Disease, CHD – Congenital Heart Defect

NYHA (New York Heart association, functional Classification)

The assessment of symptoms according to the NYHA functional class before the PPVI and 3 months after the PPVI, who managed to undergo the follow-up, was carried out in 13 patients from the ACHD Dept. and 5 patients from the Children´s Cardiac Centre, is summarized in Table 2. All 18 evaluated patients improved in NYHA and experienced improved well-being.

Table 2. NYHA functional classification before and 3 months after PPVI.

NYHA	Before the PPVI	After the PPVI
ACHD Dept.		
I	**0**	**9**
II	**9**	**3**
III	**3**	**0**
Children´s **Cardiac Centre**		
I	**5**	**better**
II	**0**	**0**
III	**0**	**0**

Notes: NYHA – New York Heart classification, ACHD Dept. – Department of Congenital Heart Disease in Adulthood, PPVI – Percutaneous pulmonary valve implantation

Evaluation of the NTproBNP

Examination of the cardiac specific marker before and after the PPVI was performed in 11 patients.

In all adult patients, there was a significant decrease until its normalization. In all patients from Children´s Cardiac Centre, NTproBNP values were still normal even before PPVI. For this reason, they did not have it examined after the procedure, and therefore they were not included in the graphic evaluation (Figure 10).

Evaluation of the change in the severity of pulmonary regurgitation

Echocardiographic evaluation of the change in the severity of PuR before and after PPVI during hospitalization was performed in all patients (Figure 2).

Evaluation of peak gradient change in a. pulmonalis before and after PPVI

Echocardiographic evaluation of the change in peak gradient in a. pulmonalis (peak gr. in mmHg) before and after PPVI during hospitalization was performed in all patients (Figure 12). The value of the peak gradient up to 3 m/s is considered to be the flow gradient in severe PR. In all patients, the peak gradient (including the flow gradient) decreased significantly after PPVI.

Parameters evaluated by MRI

All patients underwent an MRI examination before the PPVI, but only half of the patients (9 patients from ACHD Dept. and 1 patient from Children´s Cardiac Centre) were able to undergo a follow-up after 3 months after the PPVI. Figure 13-15 show the change in indexed endiastolic, endsystolic volume and RV

Figure 10. Change in NTproBNP before and after PPVI.

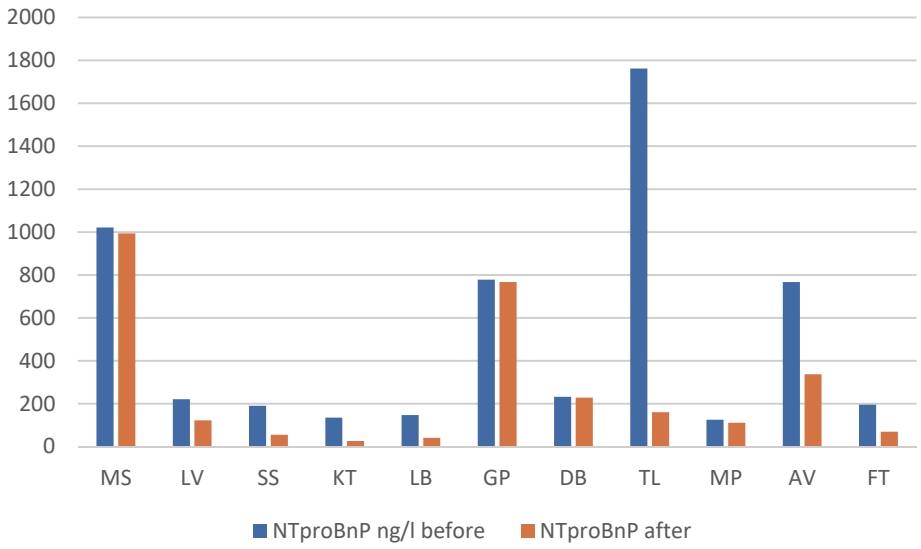
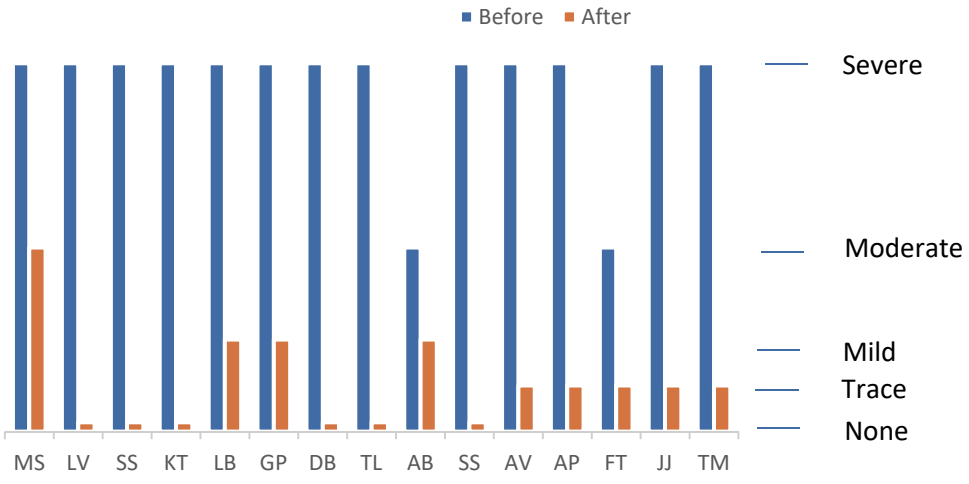


Figure 11. Change in the severity of pulmonary regurgitation before and after the PPVI.



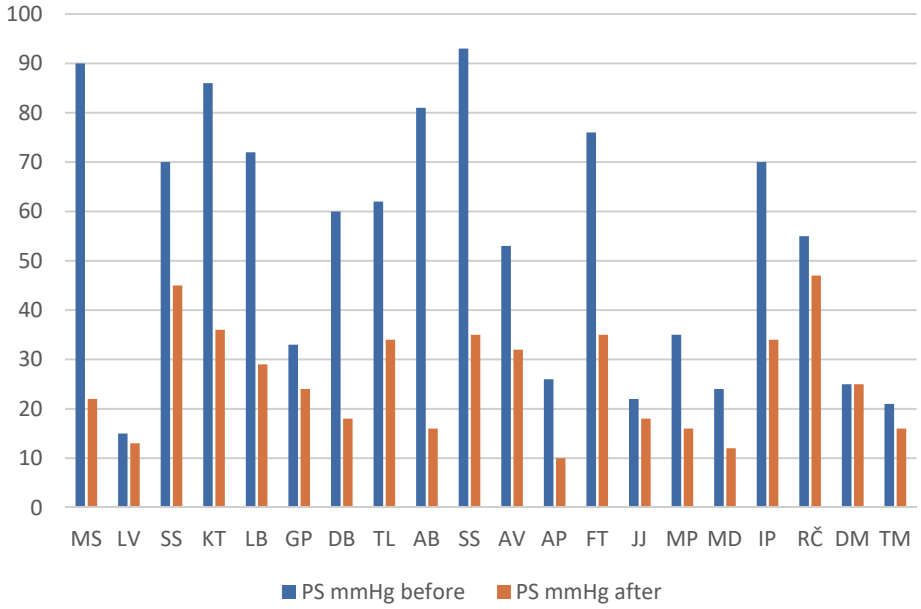


Figure 12. Change in peak gradient in a. pulmonalis before and after PPVI.

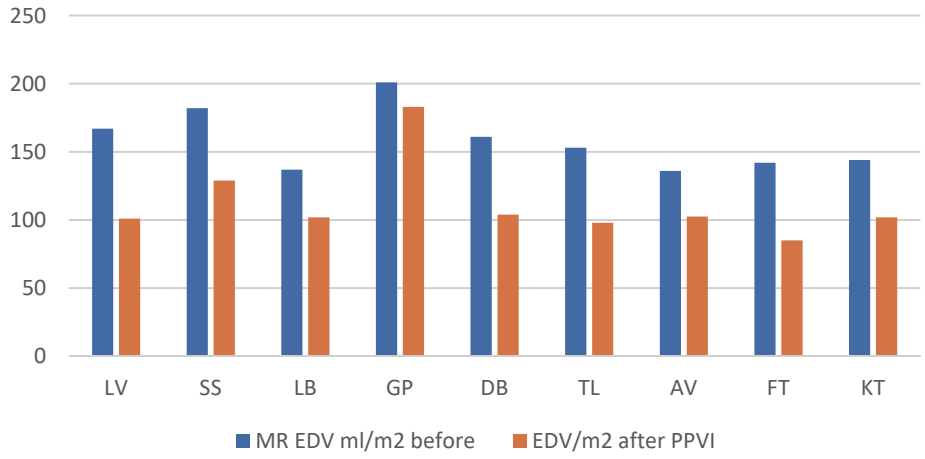


Figure 13. Change in indexed endiastolic volume of RV before and after PPVI.

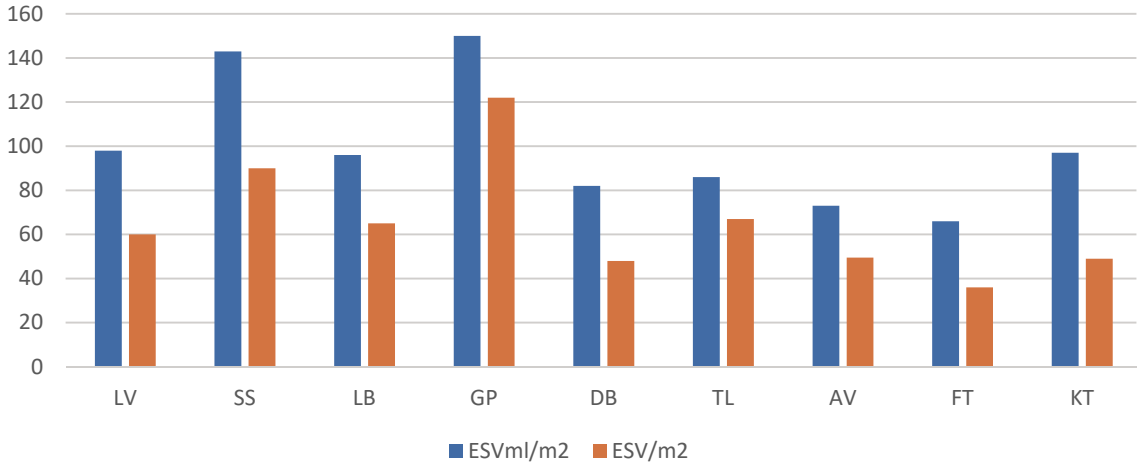


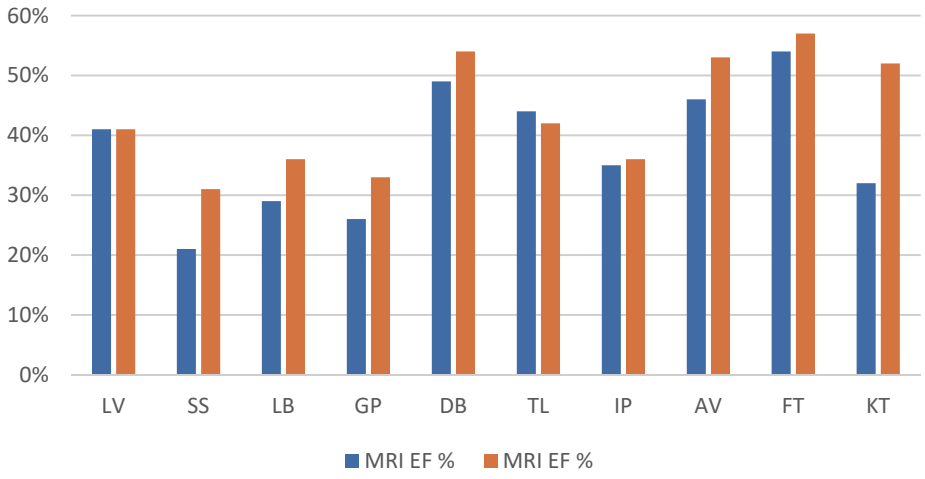
Figure 14. Change in indexed endsystolic RV volume before and after PPVI.

function before and after PPVI. In all patients, there was a significant regression of indexed RV volume at the end of diastole and at the end of systole. RV function improved in 6 patients and remained unchanged in 4 patients.

Complications

Periprocedural complications (stent fracture, coronary artery oppression) were not recorded. One patient suffered from a deflation disorder and balloon rupture, which we assess as a rare technical complication related to instrumentation. In one patient, repeated surgical in-

Figure 15. Change in the ejection fraction of the RV before and after the PPVI.



tervention of vascular access was required, which is related to the fragility of the vascular wall in CHD. Late complications - IE, death and the need for further reintervention have not been recorded so far.

Discussion

The survival of patients with CHD into adulthood is increasing, which is conditioned by several factors, but especially by constantly improving treatments (13). According to available statistical data in Slovakia registered in the NCZI (14) , 31.6% of live births accounted for newly diagnosed children with CHD in 2021. Up to ten patients with TOF are transferred from Children' Cardiac Centre to ACHD Centre yearly. From the above, it follows that the number of patients with CHD will increase. These patients require lifelong follow-up first with a paediatric cardiologist and then with a cardiologist for adult patients, ideally with experience with CHD.

The findings of many of these patients eventually reach the need for another solution, for many it means multiple reoperations. The introduction of the PPVI in 2000 was a breakthrough (7) because in suitable candidates it fully replaced its surgical implantation (15). The historically first PPVI procedure in Slovakia was carried out in June 2021 at our workplaces in National Institute of Cardiovascular Diseases – the ACHD Department and the Department of Functional Diagnostics of Children´s Cardiac Centre. After a thorough selection of patients based on the ESC Guidelines for CHD (3), 20 patients were successfully implanted at our workplace by the end of 2024. Two types of bioprostheses were used (Edwards Sapien and Venus P). The PPVI program has been successfully launched in the Slovak Republic and the findings of other patients are actually at the stage of assessing the suitability of metric parameters.

The most common diagnosis for which PPVI is indicated is TOF, less often patients with rarer diagnoses. Patients with TOF also dominated in our cohort, and to a lesser extent more complex CHD (16). Patients with CHD are often subjected to at least one additional reoperation, which increases the risk of perioperative complications and mortality (17).

Because of PPVI procedure, patients are spared reoperations. In our cohort, at least 6 patients were reoperated once, of which 1 patient up to five times, i.e. more than half of our patients prevented another reoperation.

Objective assessment of clinical relevant symptoms and inclusion in the functional class of NYHA, mostly young patients with CHD, remains a challenge. The symptom of shortness of breath is often ambiguous in them. On one hand, these are young patients with a sufficient functional reserve who regularly do sports. On the other hand, on the contrary, they are untrained, or do not do sports even recreationally, and purposefully save themselves. In the comprehensive assessment of a patient with TOF, ergometry can be very useful and requires clinical experience (18). According to the ESC Guidelines for ACHD 2020 (24), symptomatic patients are indicated for RVOT in class I. However, several studies have confirmed increased mortality in asymptomatic patients with TOF who, despite meeting the indication criteria, did not undergo RVOT intervention (19). In all patients, we showed improvement in the functional class of NYHA, and most admitted after 3 months that they could not even imagine that they would breathe better.

Another parameter monitored was the change in the NTproBNP level. In young patients, including children with CHD, this value is often in the normal range, or only slightly elevated to 300 ng/l. A significantly increased value of this marker clearly indicates the progression of RV failure, regardless of age 20. This fact has also been proven in the case of our 21-year-old. a patient whose NTproBnP level was 1800ng/l before the PPVI and decreased to 150ng/l after 3 months.

In the majority of patients, the main indication for PPVI was PuR, as a natural consequence of CHD correction. Over time, this leads to volume overload to the achievement of volumetric MRI criteria, which, in addition to assessing the clinical condition, are decisive in deciding on subsequent intervention (3).

In all patients in the cohort, we recorded a significant change in PuR due to PPVI, which was also reflected in the regression of indexed RV volumes.

In some patients, the main indication for PPVI was RVOT stenoinsufficient with documented RV dysfunction. In the case of stenoinsufficiency RVOT, the volumetric criteria are often not met, or are only borderline. In these patients, we tend to encounter a decrease in RV function. This results by analogy from unfavorable hemodynamics, when RV expels blood into the lungs through a narrowed, degeneratively altered conduit. The timing of the procedure indication itself must be carefully considered, as it is not always easy to follow volumetric criteria, each patient is individual. Thus, the goal of a well-timed indication is ideally to achieve reverse remodeling and prevent premature mortality. This was demonstrated in the Indicator study cohort, the later patients were treated, the worse their survival was (21).

Complications of PPVI (22, 23) are divided into early (e.g. stent fracture, coronary artery oppression) or late (e.g. IE, need for further reintervention). These were not recorded in our study population.

However, there were rarer, mostly technical complications related to the instrumentation (deflation disorder and balloon burst). In one patient, repeated surgical intervention of vascular access was required, which is related to the fragility of the vascular wall.

All patients are indicated after PPVI for dual antiplatelet therapy for 6 months, then lifelong acetylsalicylic acid monotherapy. Discontinuation of acetylsalicylic acid, as a factor in the development of thrombosis and IE, was demonstrated in a prospective study on a sample of 86 patients (24). Last but not least, it should be noted that prophylaxis and prevention of IE are indicated for life in all these patients, as they are at high risk (9).

The aim of the PPVI is to prevent irreversible failure of the RV, in the most ideal case to achieve its so-called reverse remodeling, which we have also demonstrated in our pilot study.

However, it should be noted that the bioprosthesis will also degenerate over time. Another interventional option is the subsequent interventional procedure of implantation of a bioprosthesis into the bioprosthesis - „valve in valve“.

Conclusion

PPVI has become an accepted and integral part of the solution of RVOT dysfunction not only in patients with TOF, but also in less common CHD. In suitably selected candidates, it has proven to be a safe, effective, especially less invasive and gentler alternative to surgical solutions. PPVI became implemented by our skilled team into common practice also in our country. If the patient is not a suitable candidate for this procedure, he/she is indicated for classic cardiac surgery. Given the specificity of the CHD issue, the centralization of these patients is crucial. In our country, this principle has been applied for more than a decade and patients with complex CHD are managed at single specialized centre – our ACHD Dept., which has been granted the status of an expert workplace by the Ministry of Health of the Slovak Republic. All patients with CHD even those after PPVI, require lifelong follow-up at such a centre in cooperation with a regional cardiologist and other specialists.*

*__Compliance with Ethics Requirements:__ Authors declare no conflict of interest regarding this article. The authors declare, that all the procedures and experiments of this research respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2008 (5), as well as the national law.

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