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Case Report

Description of Mechanical Valves Use in Pediatric Patients: A Case Series and Literature Review

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Abstract

Valvular disease carries high morbidity and mortality rates in the pediatric population, and initial treatment should aim to repair the valve. However, there are cases where repair is not feasible, so valve replacement is considered. There are several options for aortic valve replacement including the Ross procedure, mechanical prostheses, and biological prostheses. For mitral valve replacement, options are limited to mechanical prostheses and a few emerging alternatives. This article describes 6 patients who underwent valve replacement; two had mitral valve replacement and four had aortic valve replacement. In all cases, mechanical prostheses were used, resulting in satisfactory outcomes during follow-up. Additionally, a literature review is presented, concluding that for aortic valve replacement, the Ross procedure has lower mortality rates than mechanical prostheses but higher reintervention rates. For mitral valve replacement, therapeutic options are limited, and mechanical prostheses continue to be an acceptable alternative.

Keywords: Aortic valve replacement, Mitral valve replacement, Pediatrics, Children, Mechanical prosthesis

Introduction

Congenital and acquired valvular disease represents high morbidity and mortality rates in the pediatric population, particularly in developing countries. Treatment should initially focus on repair, and when valvuloplasty fails, valve replacement should be considered. For aortic valve replacement (AVR), there are various options, including the Ross procedure (pulmonary autograft in the aortic position and allograft in the pulmonary position), mechanical prostheses, biological prostheses, and homografts. Each option offers different advantages and limitations, and there is no ideal choice as evidence remains controversial.

In terms of mitral valvulopathy, numerous repair techniques exist. However, when these are not successful and replacement becomes necessary, options are limited, leaving the mechanical prosthesis as the preferred alternative, considering the risks associated with lifelong anticoagulation and possible reoperation due to somatic growth in children.

This article aims to describe the outcomes of aortic and mitral valve replacements in pediatric populations and to review current literature to support and contrast the decisions made regarding the approaches taken.

Case Series

Case 1

A 21-month-old patient presented with a 16-day history of fever associated with respiratory distress, leading to antibiotic treatment for suspected pneumonia. However, the patient developed left hemiparesis and generalized edema, deteriorating respiratory function requiring

intubation. A systolic murmur was detected on physical examination, and an echocardiogram revealed a vegetation on the mitral valve's posterior leaflet and severe mitral insufficiency. Blood cultures were positive for Staphylococcus haemolyticus, and antibiotic management included meropenem, vancomycin, and amphotericin. Despite medical management with inotropes and antibiotics, the patient developed refractory cardiogenic shock, leading to the decision for surgical intervention for valvuloplasty or mechanical valve replacement. During surgery, the mitral valve was found to be severely insufficient, with extensive damage to the posterior leaflet, a 2x2 cm vegetation, and perforation throughout the valve, resulting in severe insufficiency, fragility, and damage extending to the posterior annulus. These findings necessitated opting for a mechanical aortic prosthesis, placed in a supra-annular mitral position due to heart size, ensuring an adequately sized valve to avoid short-term replacement. There were no complications post-bypass, and the native mitral valve was sent for analysis, confirming colonies of MRSA. Post-operative management included antibiotics trimethoprim-sulfamethoxazole and daptomycin. Early postoperative follow-up showed good biventricular function with satisfactory prosthetic function and reduced pulmonary pressure. The patient was discharged on day 52 of hospitalization with an INR of 3.5. Follow-up indicated good recovery of left hemibody movements without further symptoms.

Case 2

This concerns a 23-month-old patient who had been followed up in outpatient care because since the age of 3 months, the patient had experienced multiple hospitalizations due to respiratory conditions, managed with mechanical ventilation in infancy and other non-invasive treatments without sequelae. Additionally, the patient exhibited poor growth associated with diaphoresis during feeding. Therefore, an echocardiogram was performed, revealing dilated cardiomyopathy associated with mitral valve insufficiency and valvular dysplasia, leading to deterioration of ventricular function. Medical management commenced with enalapril, furosemide, carvedilol, spironolactone, and beta-methyldigoxin, considering the patient as having dilated cardiomyopathy secondary to mitral valve insufficiency due to valvular dysplasia, possibly parachute mitral valve. Following this, an MRI was performed to rule out non-compacted myocardium and evaluate the possibility of surgical management. Upon ruling out non-compacted myocardium, a medical-surgical board discussed the options of waiting, performing valvuloplasty, or proceeding with mechanical valve replacement. Given the deterioration of ventricular function and the onset of the COVID-19 pandemic, it was decided to proceed with mitral valvuloplasty, and if not possible, to replace with a mechanical prosthesis. After making this decision, while waiting to schedule the surgery, the patient presented with respiratory distress associated with signs of respiratory difficulty, leading to an emergency consultation where the patient was considered to be experiencing acute heart failure. Therefore, given the hospital admission due to cardiac illness, the decision was made to perform surgery during this hospital stay. During surgery, the mitral valve exhibited severe dysplastic changes, with retraction of the subvalvular apparatus and calcification of the tendinous chords. This made valvuloplasty impossible, so the anterior leaflet was resected, and the posterior leaflet was left in place to allow for the implantation of the subvalvular apparatus, where a 23 mm inverted mechanical aortic prosthesis was implanted. There were no complications following the exit from extracorporeal circulation. Postoperatively, echocardiography showed proper function of the mechanical prosthesis without leaks and good systolic function of the left ventricle without pulmonary hypertension. Consequently, the patient was discharged on hospital day number 37, and during the 2-month post-replacement follow-up, the patient's family reported improvement in diaphoresis during feeding and maintained therapeutic INR levels without any other symptoms.

Case 3

This case involves a 12-year-old male patient with a diagnosis of congenital

aortic stenosis due to a bicuspid aortic valve, identified when he was 6 months old. He had been under periodic follow-up with pediatric cardiology. Symptoms of functional class deterioration and chest pain emerged, prompting an echocardiogram that revealed severe aortic stenosis with concentric left ventricular hypertrophy. The decision was made to proceed with aortic valvuloplasty using a balloon, which failed due to severe deformity of the valvular plane that prevented retrograde passage of the guide wire into the left ventricle. Given this situation and considering the severity of the lesion, surgical commissurotomy of the aortic valve was indicated and performed without complications. Following this, severe aortic insufficiency developed, characterized by a dilated left ventricle with signs of volume overload and good systolic function, and mild pulmonary hypertension. Congestive management was initiated with captopril, spironolactone, and furosemide, and the patient was scheduled for aortic valve replacement (AVR) with a mechanical prosthesis, where valve dysplasia, thickening, and retraction were found. A 21 mm ON-X mechanical valve was implanted. There were no complications during the exit from extracorporeal circulation, and the postoperative echocardiogram showed a normally functioning mechanical prosthesis, concentric left ventricular hypertrophy, and good biventricular function. The patient was discharged on hospital day number 35 with a therapeutic INR. At the 10-year follow-up, the patient showed good progress with no chest pain or syncope, and a New York Heart Association (NYHA) functional class I. The echocardiogram reported a normally sized left ventricle, normal global and segmental contractility with an ejection fraction (EF) of 67%, and a well-implanted aortic mechanical prosthesis with normal movements and an effective area of 1.7 cm^2 without pulmonary hypertension. However, due to poor adherence to anticoagulation management and a subtherapeutic INR, the patient experienced an ischemic stroke, resulting in left hemiparesis, which was managed with physical therapy, leading to significant improvement in the paresis without resulting in disability. Following this event, the patient resumed anticoagulation management and has remained within therapeutic INR ranges without further complications.

Case 4:

This concerns a 10-year-old patient with a history of congenital aortic stenosis, managed with aortic valvuloplasty using a balloon at 45 days of life. The patient later presented to emergency care with progressive dyspnea on moderate to minimal exertion, considered to be due to decompensated heart failure and community-acquired pneumonia. Congestive and antibiotic management were initiated, and echocardiography revealed severe aortic insufficiency associated with moderate stenosis impacting the left ventricle due to dilation and dilation of the ascending aorta. It was determined that the patient would benefit from aortic valve replacement (AVR) surgery using either a mechanical prosthesis or homograft. However, due to the unavailability of a homograft, a mechanical prosthesis was chosen. The procedure revealed a bicuspid aortic valve due to congenital fusion of the right and left commissures with retracted and thickened coaptation edges, without calcification or vegetations, and a healthy valvular ring where a 21 mm ON-X mechanical prosthesis was implanted without complications. Following the exit from extracorporeal circulation, the patient exhibited supraventricular arrhythmia due to extrasystole with bigeminy, leading to the initiation of metoprolol treatment and evaluation by electrophysiology, which concluded that the patient had ventricular and atrial extrasystoles conducted secondary to valvular cardiopathy, ventricular hypertrophy, and systolic overload, and should continue treatment with a beta-blocker at the maximum tolerated dose. The postoperative echocardiogram showed a competent ON-X-21mm mechanical prosthesis with the expected gradient, a dilated left ventricle with good contractility, and no signs of pulmonary hypertension. The patient was discharged on hospital day 21 with therapeutic INR levels. At the 1-month follow-up, the patient reported improvement in dyspnea and maintained therapeutic INR levels without further complications.

Case 5

This case involves a 9-year-old patient with a history of aortic valve stenosis due to a bicuspid aortic valve, who underwent percutaneous balloon valvuloplasty in the neonatal period, followed by the establishment of severe insufficiency leading to hypertrophy and dilation of the left ventricle due to volume overload and good biventricular function, associated with deterioration of functional class. Therefore, it was decided that the patient would benefit from AVR with a mechanical prosthesis. During the procedure, valve dysplasia and failure in the architecture of the leaflets, which did not allow for coaptation, were discovered. Following this, a 21 mm ON-X mechanical prosthesis was implanted without complications. The patient showed good postoperative progress, with control echocardiography demonstrating a normally functioning prosthesis and concentric hypertrophic cardiomyopathy with preserved systolic function. During hospitalization, the patient showed good progress; however, during the titration of warfarin, the patient experienced self-limiting nasal bleeding associated with septal varices and a supra-therapeutic INR, necessitating the suspension of warfarin and the switch from nasal cannula to oxygen mask use. Following this, no further complications occurred, and the patient was discharged on hospital day 21. At the 8-month follow-up, the patient had therapeutic INR levels, reported improvement in functional class, had not experienced new bleeding episodes, and had a normally functioning prosthesis without further alterations.

Case 6

This case involves a 10-year-old patient with a history of ventricular septal defect (VSD) closure at age 5, who was referred due to a 2-year deterioration in functional class, which had been associated with febrile peaks for the past 2 months. On physical examination, the patient exhibited a hyperdynamic precordium, bounding pulses, and a systolic-diastolic murmur at the aortic focus. An echocardiogram revealed a well-anchored VSD patch without residual defect, a bicuspid aortic valve with severe insufficiency greatly impacting the left ventricle, systolic dysfunction, and an image of vegetation on the right coronary cusp. Therefore, it was considered that the symptoms were due to an unmonitored bicuspid aorta, which subsequently presented two years of insufficiency, and in the last two months, endocarditis. Blood cultures were taken, which were negative, and empirical antibiotic management and congestive management were initiated. Once the infectious process improved, an AVR was planned with either a biological prosthesis (aortic homograft) or a mechanical prosthesis replacement. During hospitalization, the patient exhibited hyperreflexia and exhaustible clonus in the lower limbs, prompting a cerebral MRI that showed a microinfarct in the left caudate nucleus head and hyperintense cortico-subcortical lesions in the left parietal and fronto-insular regions, with an evident hemorrhagic component. This led to the consideration of a cardioembolic etiology due to the infectious process, and angiography was performed to assess collateral circulation, which showed adequate perfusion. Surgery was subsequently carried out, finding left ventricular dilation with preserved contractility. The three-cusp aortic valve showed rupture of the non-coronary and left cusps' insertions with multiple vegetations on the coaptation edges of these two cusps. The VSD dacron patch was removed, and the defect was closed with continuous monofilament suturing. Postoperatively, the patient showed good progress, with an echocardiogram revealing a slight paravalvular leak from the aortic valve prosthesis and good contractility with a 60% ejection fraction without further complications. The patient was discharged on day 53 of hospitalization with therapeutic INR levels. Neurologically, the patient exhibited no sequelae or disability, but follow-up was lost, so the current situation is unknown.

Table 1: Sociodemographic data and reason for consultation

Case	**1**	**2**	**3**	**4**	**5**	**6**
Age	21 m	23m	12 a	10 a	9 a	10 a
Gender	m	m	m	f	m	M
Weight	14kg	10.6kg	40kg	30kg	27 kg	27 kg
Body Surface area	0.6 m2	0.5 m2	1.28 m2	1.08 m2	1 m2	1 m2
Reason for consultation	Fever	Failure to thrive	Dyspnea	Dyspnea	Dyspnea	Fever
Reason for replacement	Endocarditis	Mitral in parachutes	IAS post commissurotomy	IAS post balloon valvuloplasty	IAS post balloon valvuloplasty	endocartitis

Case	1	2	3	4	5	6
Background	negative	Hospitalization recurrent by pictures respiratory	VAP + CQ	VAP	VAP	CIV closure with dacron patch
Dx ECO	IMS	IMS	IAS + EAM	IAS + EAM	IAS + EAM	IAS
Initial FE	65%	60%	65%	62%	64%	45%
Control FE	68%	60%	68%	65%	65%	60%
Initial PSAP	40 mmHg	50 mmHg	26 mmHg	40 mmHg	22 mmHg	54 mmHg
Control PSAP	26 mmHg	25 mmHg	20 mmHg	25 mmHg	20 mmHg	
Initial contract vein	6.6 mm	8 mm	N/A	N/A	N/A	N/A
Initial máximum aortic echo gradient	N/A	N/A	27mmHg	55 mmHg	24mmHg	
Control Máximum aortic gradient	N/A	N/A	16 mmHg	16 mmHg	11 mmHg	
DDVI	-	-	58 mm	46 mm	40 mm	60 mm
Complications	ESC		Ischemic stroke	EV	Nasal Bleeding	ESC

VAP (percutaneous aortic valvulotomy), VSD (ventricular septal defect), CQ (commissurotomysurgical), ESC (cerebral septic embolus), Dx ECO (initial diagnosis by echocardiogram), FE(ejection fraction), PSAP (pulmonary arterial systolic pressure), SMI (severe mitral regurgitation),IAS (severe aortic insufficiency) MSE (moderate aortic stenosis), EV (ventricular extrasystole)

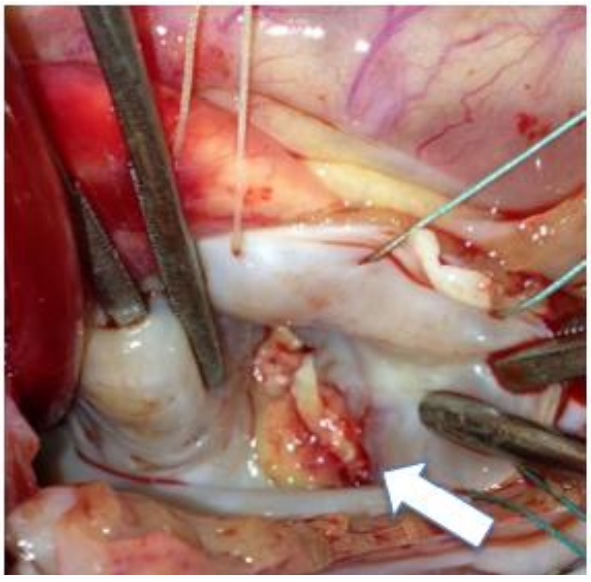

Image 1. Mitral valve endocarditis, Major involvement of the posterior mitral valve apparatus with posterior leaflet perforation (Arrow)

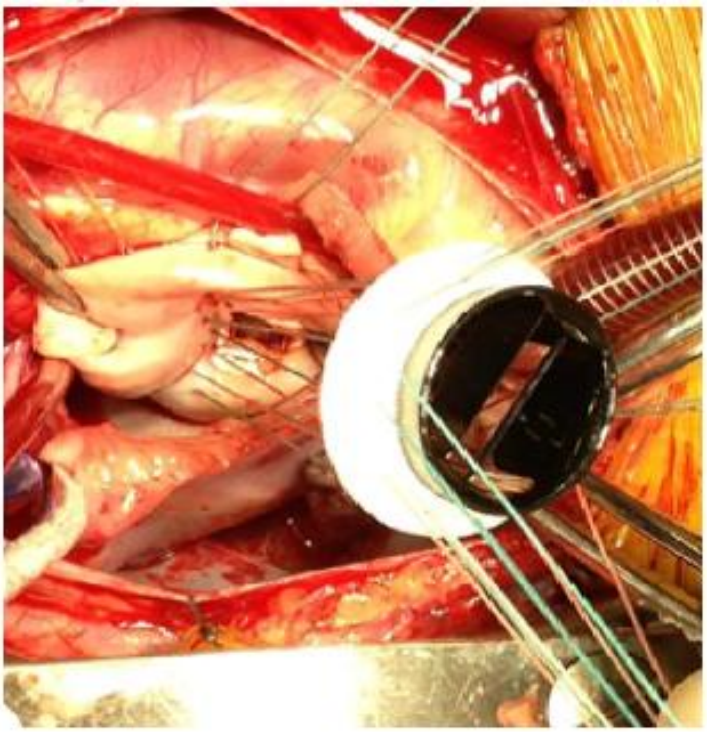

Image 2. Mitral valve replacement with mechanical aortic prosthesis

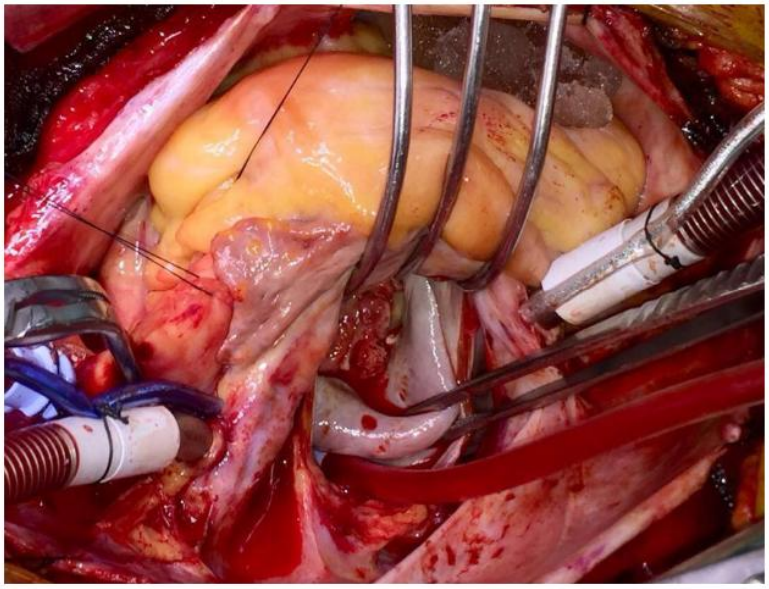

Image 3. Mitral valve in parachute. Mitral valve replacement with classical technique

Discussion

Left-sided valve replacement in pediatric patients is performed less frequently compared to adults, and systematic reviews show suboptimal results in children, reflecting the need for solutions for this patient group due to the lack of an ideal therapeutic option.

Regarding aortic replacement, early mortality in patients undergoing AVR is primarily determined by intrinsic factors such as age, the urgency of the procedure, pre-surgical hemodynamic status, and the underlying disease necessitating the replacement, rather than by the type of procedure, whether Ross or mechanical prosthesis replacement.

Late mortality in the first postoperative decade shows that the Ross procedure appears to have lower mortality compared to mechanical aortic valve replacement (MAVR), although this might be explained by selection bias as in the meta-analysis observed, most mechanical prostheses were replaced in children with rheumatic disease and connective tissue

disorders, factors that could condition higher mortality. Additionally, late mortality in patients with mechanical prostheses has decreased over time, thanks to advances in pediatric cardiovascular management. In studies where this was not the case, patients had suboptimal anticoagulation levels, a frequent issue as evidenced in one of the cases presented.

When comparing the number of reoperations, the Ross procedure and MAVR have comparable rates, but the reasons for reintervention differ. With mechanical prostheses, decoupling due to patient growth accounts for many reoperations, while the Ross surgery has the disadvantage of managing single-valve valvulopathy with double valve replacement, leaving two valves predisposed to degeneration and possible reoperation, thus resulting in a higher total number of reoperations compared to mechanical prosthesis replacement (right ventricular reoperations in Ross 4.30%/year) during the first postoperative decade, and it is expected that during the second decade, the reintervention rate will be even higher, with only 49% of patients being free from intervention for the allograft and 77% for the autograft.

The main cause of reintervention for the autograft will be aortic root dilation leading to valve regurgitation, while the main cause for the allograft will be degeneration and calcification leading to consequent valve stenosis.

This aspect must be considered when counseling parents on choosing an option for replacement. It is also worth mentioning that repairs at the right ventricular level represent the majority of these interventions and are more frequent in patients who underwent the Ross procedure during the neonatal and infancy stages.

Regarding complications, evidence clearly shows that replacement with mechanical prostheses is associated with a higher risk of thromboembolic events compared to the Ross procedure (0.76%/year vs. 0.22%/year), and the rate of endocarditis is similar between Ross procedure and MAVR.

As for mitral valvulopathy, reconstruction or repair is the most favored management in pediatric patients due to the lack of prostheses of adequate size for their position and because it avoids the use of foreign material requiring anticoagulation. Repair using autologous pericardium has been described, showing adequate endothelialization and low rates of infection and thrombosis. Despite these repair techniques, there are cases like those presented where valvuloplasty is not possible, and replacement with a mechanical prosthesis is necessary due to the lack of long-lasting options, a reality that has not changed over the past 20 years. Hence, the use of inverted aortic prostheses in the mitral position has been reported.

Conclusions

Both the Aortic Valve Replacement with a Mechanical Prosthesis (AVR-MP) and the Mitral Valve Replacement with a Mechanical Prosthesis (MVR-MP) are viable, reproducible, and safe options in both the short and long term for pediatric patients requiring valve replacement. For AVR, a case-by-case approach should be adopted, taking into account the patient who will undergo the replacement and considering the various advantages offered by other available options. For MVR, there are not many options available, hence more studies are needed to compare emerging technologies to provide better counseling and determine the best option for each patient.

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