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Research Article

Mechanical aortic valve replacement in children

Çocuklarda mekanik aort kapak replasmanı

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Abstract

Aim: Aortic stenosis is a congenital heart disease characterized by the narrowing of the aortic valve, the valve that regulates blood flow from the heart's left ventricle to the aorta. While aortic stenosis can affect individuals of all ages, including children, it poses unique challenges in pediatric patients. The severity of aortic stenosis in children can vary widely, ranging from mild to severe, and may present with symptoms such as chest pain, fatigue, and shortness of breath. If left untreated, aortic stenosis can lead to significant complications and negatively impact a child's overall health and quality of life.

Material and methods: Between February 2019 and June 2023, 38 patients were operated due to aortic valve pathologies in our hospital. Aortic valve repair was performed in 11 of these patients, and aortic valve replacement was performed in 27 patients. Patients' age, gender, body weight, aortic valve pathology, etiology of aortic valve pathology (congenital, rheumatic, infective endocarditis), presence of Marfan Syndrome, previous operation history, aortic annulus diameter, type of valve used, valve size, type of root enlargement if performed, cardiopulmonary bypass duration, cross-clamp duration, duration of intensive care unit stay, duration of ward stay, inotrope requirement, duration of inotrope use, mechanical ventilation duration, volume of drainage and mortality were retrospectively searched from patient files and hospital database.

Results: The median age of the patients was 12.96±3.38 (IQR=11.00-16.00) years, their weights ranged between 43.81±14.21 kilograms. Eight patients were female (29.6%) and 19 patients were male. (70.4%) The diagnosis was aortic stenosis in 8 patients (29.6%), aortic insuffiency in 9 patients (33.3%) and both aortic stenosis and insuffiency in 10 patients.(37%) The aortic annulus diameters of the patients were 21.59±4.64 mm. Anterior or posterior root enlargement was performed in 11 patients (40.7%) due to narrow aortic annulus. The Nick procedure was applied to 7 patients (63.6%), the Manoughian procedure to 2 patients (18.2%), and the Konno procedure to 2 patients (18.2%). Considering the mechanical aortic valve dimensions used in the patients, 5 patients had size 19 prosthetic aortic valve (18.5%), 8 patients had size 21 prosthetic aortic valve (29.6%), 8 patients had size 23 prosthetic aortic valve (29.6%), and 6 patients had size 25 prosthetic aortic valve (22.2%) were used. Mortality was observed in 3 patients.(11.1%) Causes of mortality can be listed as low cardiac output, neurological events and sepsis.

Conclusion: The ultimate goal is to ensure that children who undergo aortic valve replacement can lead healthy and fulfilling lives. By continually refining our approaches and learning from each case, we can make significant strides in the treatment of aortic valve issues in children and offer them the best possible outcomes.

Aortic valve replacement in children requires a multidisciplinary approach, with a focus not just on the surgical procedure itself but also on long-term management and support. With ongoing advancements and a collaborative mindset, we can continue to improve the care provided to these young patients and help them thrive.

Keywords: aortic valve replacement, aortic stenosis, aortic regurgitation, aortic root enlargement

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Öz

Amaç: Aort darlığı, kalbin sol ventrikülünden aorta kan akışını düzenleyen kapak olan aort kapağının daralması ile karakterize doğuştan bir kalp hastalığıdır. Aort darlığı çocuklar da dahil olmak üzere her yaştan bireyi etkileyebilirken, pediatrik hastalarda benzersiz zorluklar ortaya çıkarır. Çocuklarda aort darlığının şiddeti, hafif ile şiddetli arasında geniş ölçüde değişebilir ve göğüs ağrısı, yorgunluk ve nefes darlığı gibi semptomlarla kendini gösterebilir. Aort darlığı tedavi edilmezse önemli komplikasyonlara yol açabilir ve çocuğun genel sağlığını ve yaşam kalitesini olumsuz etkileyebilir.

Gereç ve Yöntemler: Şubat 2019-Haziran 2023 tarihleri arasında hastanemizde aort kapak patolojileri nedeniyle 38 hasta ameliyat edildi. 11 hastaya aort kapak tamiri, 27 hastaya aort kapak replasmanı yapıldı. Hastaların yaşı, cinsiyeti, vücut ağırlığı, aort kapağı patolojisi, aort kapağı patolojisinin etiyolojisi (konjenital, romatizmal, enfektif endokardit), Marfan Sendromu varlığı, geçirilmiş ameliyat öyküsü, aort anulus çapı, kullanılan kapak tipi, kapak boyutu, yapıldıysa aortik kök genişletme prosedürü, kardiyopulmoner baypas süresi, kros klemp süresi, yoğun bakımda kalış süresi, serviste kalış süresi, inotrop gereksinimi, inotrop kullanım süresi, mekanik ventilasyon süresi, drenaj miktarı, revizyon gereksinimi ve mortalite retrospektif olarak hasta dosyalarından ve hastane veri tabanından tarandı.

Bulgular: Hastaların medyan yaşı 12,96±3,38 (IQR=11,00-16,00) yıl olup, ağırlıkları 43,81±14,21 kg arasında değişmekteydi. Sekiz hasta (%29.6) kadın, 19 hasta erkekti(%70.4). 8 hastada (%29.6) aort darlığı, 9 hastada (%33.3) aort yetmezliği ve 10 hastada (%37) hem aort darlığı hem de yetmezlik tanısı kondu. Hastaların aort anulus çapları 21,59±4,64 milimetre idi. 11 hastada (%40.7) dar aort anulus nedeniyle anterior veya posterior kök genişletmesi yapıldı. 7 hastaya Nick (%63.6), 2 hastaya Manoughian (%18.2) ve 2 hastaya Konno (%18.2) prosedürü uygulandı. Hastalarda kullanılan mekanik aort kapak ölçülerine bakıldığında 5 hastada 19 numara protez aort kapağı (%18.5), 8 hastada 21 numara protez aort kapağı (%29.6), 8 hastada 23 numara protez aort kapağı (%29.6), 6 hastada ise 25 numara protez aort kapağı (%22.2) kullanıldı. Mortalite 3 hastada (%11.1) görüldü Mortalite nedenleri düşük kardiyak output, nörolojik olaylar ve sepsis olarak sıralanabilir.

Tartışma: Nihai hedef, aort kapağı replasmanı yapılan çocukların sağlıklı ve tatmin edici bir yaşam sürdürebilmelerini sağlamaktır. Yaklaşımlarımızı sürekli iyileştirerek ve her vakadan öğrenerek, çocuklarda aort kapağı sorunlarının tedavisinde önemli adımlar atabilir ve onlara mümkün olan en iyi sonuçları sunabiliriz.

Sonuç: Çocuklarda aort kapak replasmanı, sadece cerrahi prosedürün kendisine değil, aynı zamanda uzun vadeli yönetim ve desteğe odaklanan multidisipliner bir yaklaşım gerektirir. Devam eden ilerlemeler ve işbirlikçi bir zihniyetle, bu genç hastalara sağlanan bakımı iyileştirmeye ve gelişmelerine yardımcı olmaya devam edebiliriz.

Anahtar Kelimeler: Aortik kapak replasmanı, Aort darlığı, Aort yetmezliği, Aortik kök genişletme

Introduction

Aortic stenosis is a congenital heart disease characterized by the narrowing of the aortic valve, the valve that regulates blood flow from the heart's left ventricle to the aorta. While aortic stenosis can affect individuals of all ages, including children, it poses unique challenges in pediatric patients. The severity of aortic stenosis in children can vary widely, ranging from mild to severe, and may present with symptoms such as chest pain, fatigue, and shortness of breath. If left untreated, aortic stenosis can lead to significant complications and negatively impact a child's overall health and quality of life.

Aortic valve replacement (AVR) is a surgical procedure commonly employed to treat severe cases of aortic stenosis in children. The primary goal of AVR is to relieve the obstruction and restore normal blood flow through the aortic valve. Over the years, advancements in medical technology and surgical techniques have significantly improved the outcomes of aortic valve replacement in children, leading to better long-term prognosis and enhanced quality of life.

Despite the promising results in aortic valve repair, AVR is needed in severely deformed valves secondary to repetitive interventions and repairs.[1] Although mechanical valves are also available in small numbers, they can still be used from certain age group patients.[2] These mechanical prostheses are not suitable for use in infants and young children. In these patients, aortic root enlargement methods can be used in the presence of narrow aortic annulus. Posterior root enlargement methods can be listed as Nick and Manoughian. Anterior root enlargement method is Konno procedure.[3]

With these methods, it may be possible to use larger prostheses. The use of mechanical prosthesis also reveals the need for lifelong anticoagulant use. Anticoagulant use in children is a very complicated issue.

In addition, a female patient needs adjustment due to pregnancy in the following periods. Valve deformation that develops after biological valve replacement is quite rapid in children compared to adults. This is caused by an increased immunological response and increased calcium metabolism in young patients.

Although aortic homografts are a suitable alternative for aortic root reconstruction in the pediatric population, difficulties in obtaining homografts limit this use.[4]

In our study, we examined the results of patients who underwent AVR due to aortic valve pathologies. By elucidating the current knowledge and advancements in this field, this paper aims to contribute to the existing body of literature and provide valuable insights for clinicians, researchers, and families seeking comprehensive information on aortic stenosis and aortic valve replacement in children.

Materıal and Methods

Between February 2019 and June 2023, 38 patients were operated due to aortic valve pathologies in our hospital. Aortic valve repair was performed in 11 of these patients, and AVR was performed in 27 patients. All patients who were younger than 18 years of age at the time of the operation and who had undergone prosthetic AVR were included in the study. Patients who were older than 18 years of age at the time of the operation and who had more than one valve replacement were excluded from the study.

Patients' age, gender, body weight, aortic valve pathology, etiology of aortic valve pathology (congenital, rheumatic, infective endocarditis), presence of Marfan Syndrome, previous operation history, aortic annulus diameter, type of valve used, valve size, type of root enlargement if performed, cardiopulmonary bypass (CPB) duration, cross-clamp (CC) duration, duration of intensive care unit (ICU) stay, duration of ward stay, inotrope requirement, duration of inotrope use, mechanical ventilation duration, amount of drainage , need of revision and mortality were retrospectively searched from patient files and hospital database.

Warfarin sodium and enoxaparin were given as anticoagulants to all patients who underwent mechanical valve replacement. The international ratio (INR) target was kept between 2.0-3.0. In this process, patients and families were given warfarin sodium use training, and patients were called for follow-up at close intervals in the first 6 months to assess compliance.

Early mortality is the mortality that develops within the first month after the operation. Mortality was also controlled from the national health screening system in addition to the hospital database.

The follow-up period was calculated as the time between the operation date and the last hospital admission.

Operative Procedure: All patients were operated with median sternotomy and CPB at 28 degrees hypothermia. In all patients, del-Nido cardioplegia was administered antegradely via an aortic root needle and by direct coronary perfusion from the coronary ostia after aortotomy. All aortic valves were mounted in the annular position with individual plegitic sutures. Aortic root enlargement was performed in 11 patients to implant larger valves. The type of root enlargement performed was the Nick procedure in 7 patients, the Manoughian procedure in 2 patients, and the Konno procedure in 2 patients.

Statistical Analysis

Continuous data are presented as Mean ± SD, Median (IQR), whereas categorical data are presented as frequency (n) and percentage (%). Normality was tested using the Shapiro–Wilk test. All statistical analyses were performed using IBM SPSS (Statistical Package for the Social Sciences) Statistics ver.25.

Results

The median age of the patients was 12.96±3.38 (IQR=11.00-16.00) years, their weights ranged between 43.81±14.21 kilograms. Eight patients were female (29.6%) and 19 patients were male.(70.4%) The diagnosis was aortic stenosis in 8 patients (29.6%) and aortic insuffiency in 9 patients (33.3%) and both aortic stenosis and insuffiency in 10 patients(37%).

Considering the etiology of aortic valve pathology, rheumatic valve disease was observed in 5 patients (18.5%), congenital valve disease was observed in 19 patients (70.4%), and infective endocarditis was observed in 3 patients (11.1%). When congenital valve pathologies were examined, bicuspid aortic valve was seen in 16 patients (84.2%), while other etiologies were subaortic membrane, opera transposition of great arteries and truncus arteriosus, and a total of 3 patients (15.9%). 4 patients had Marfan syndrome (14.8%). Fifteen patients (55.6%) were patients who had undergone previous cardiac surgery and were reoperated.

The aortic annulus diameters of the patients were 21.59±4.64 milimeters. Anterior or posterior root enlargement was performed in 11 patients (40.7%) due to narrow aortic annulus. The Nick (63.6%) procedure was applied to 7 patients, the Manoughian (18.2%) procedure to 2 patients, and the Konno procedure (18.2%) to 2 patients.

Considering the mechanical aortic valve dimensions used in the patients, 5 patients had size 19 prosthetic aortic valve

(18.5%), 8 patients had size 21 prosthetic aortic valve (29.6%), 8 patients had size 23 prosthetic aortic valve (29.6%), and 6 patients had size 25 prosthetic aortic valve. (22.2%) were used.

All patients were operated under CPB and the Mean CPB time was 155.37±56.05 minutes, CC time was 107.78±43.65 minutes. Inotropic support was started in all patients after CPB. Depending on the hemodynamic status of the patients, inotropes continued during the intensive care period. The duration of inotrope use was 64.81±96.07 (IQR=16.00-48.00) minutes.

The follow-up period of the patients on mechanical ventilator was 67.70±148.27 hours. Patients who continued to have hemodynamic instability and could not wean from CPB or needed CPB again after the surgery were transferred to the intensive care unit under extracorporeal membrane oxigenarator(ECMO) support. In patients who could leave ECMO according to hemodynamic status, weaning was performed at the bedside in the ICU. The patients who needed ECMO were 2 (7.4). Three patients (11.1%) were re-operated on the first day after surgery due to bleeding. The drainage volume of the patients in the first 24 hours was measured as 646.30±245.31 mililiters.

Mortality was observed in 3 patients (11.1%) Causes of mortality can be listed as low cardiac output, neurological events and sepsis.

The patients were followed up in the pediatric cardiovascular surgery intensive care unit after AVR surgery. The patients were taken to the ward after a few days. The hospitalization period in the pediatric CVS ICU was 5.00±6.89 days, and the hospitalization period in the ward was 10.30±7.05 days. The follow-up period of the patients was 15.04±16.99 (IQR=1-28) days.

Discussion

AVR in children is quite a complex procedure. It involves replacing a damaged or malfunctioning aortic valve with a prosthetic valve. It's often necessary when the aortic valve doesn't function properly, leading to conditions such as aortic stenosis or regurgitation. However, performing this surgery in children poses unique challenges compared to adults. One of the primary challenges is selecting the appropriate prosthetic valve for children. Children's hearts are still growing, which means that a valve that is initially well-fitted may become too small over time. Surgeons need to consider the child's age, size, and expected growth to choose a valve that can accommodate their future needs [5,6].

Additionally, the age of the child plays a significant role in the decision-making process. Infants and young children require specialized care due to their smaller anatomy and higher surgical risks. Sometimes, aortic valve repair may be attempted before considering replacement, particularly in cases where the valve structure can be preserved [7].

Aortic valve repair is an excellent alternative when possible because it preserves the patient's own valve tissue, minimizing the need for long-term anticoagulation therapy. However, repair isn't always feasible, especially in severe cases or when the valve damage is extensive [8].

Another critical aspect is the timing of the surgery. In some cases, AVR in children is performed as an emergency procedure due to severe symptoms or life-threatening complications. However, when possible, surgeons may choose to delay the surgery to allow the child to grow, minimizing the need for multiple surgeries as they age [9].

Post-operative care is crucial in ensuring the success of the procedure. Children may require close monitoring in the intensive care unit and may need to be on various medications to manage pain, prevent infections, and regulate heart function. Cardiac rehabilitation and follow-up visits are also essential to monitor the child's progress and ensure proper healing. Long-term outcomes and quality of life are also important considerations [10].

While AVR can greatly improve a child's condition and quality of life, it's crucial to remember that prosthetic valves have limited durability. Children may require additional surgeries or interventions as they grow to replace the valve as they outgrow the initial implant.This situation highlights the importance of comprehensive follow-up care, including regular echocardiograms, to assess the function of the prosthetic valve and monitor any potential complications. Close collaboration between cardiologists, surgeons, and the child's family is essential to ensure the best long-term outcomes [11].

In recent years, there have been advancements in the field of AVR in children. Minimally invasive techniques and the development of newer prosthetic valves have improved outcomes and reduced the invasiveness of the procedure. Ongoing research aims to further refine surgical techniques and develop more durable and growth-friendly prosthetic valves for children [12].

However, lifelong anticoagulant use is required in prosthetic valve replacement. There are handicaps in the use of anticoagulants in children. These can be listed as increased

risk of bleeding, difficult dose adjustment, monitoring requirements, limited data on safety and efficacy, potential interaction with other medications, lifestyle restrictions and psychological impact [13].

The field of pediatric cardiac surgery is continuously evolving, and with advancements in medical technology, we can provide better outcomes for children with aortic valve issues. Collaborative efforts between surgeons, researchers, and healthcare professionals are vital to improving the surgical techniques and long-term care for these young patients

Conclusion

In conclusion, the ultimate goal is to ensure that children who undergo aortic valve replacement can lead healthy and fulfilling lives. By continually refining our approaches and learning from each case, we can make significant strides in the treatment of aortic valve issues in children and offer them the best possible outcomes.

Aortic valve replacement in children requires a multidisciplinary approach, with a focus not just on the surgical procedure itself but also on long-term management and support. With ongoing advancements and a collaborative mindset, we can continue to improve the care provided to these young patients and help them thrive.

Ethical Approval

The Ankara Bilkent City Hospital Clinical Researches Ethics Committee, (No: E2-23-3988, Date: 25/04/2023) has authorized all techniques used in this work. The authors declare that they adhered to the ethical norms of the 1975 Helsinki Declaration, as revised in 2008.

Conflict of Interest

The authors declare no conflict of interest.

Disclosure

None.

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