

THE EFFECTS ON PULMONARY ARTERY PRESSURE OF EARLY SURGICAL REPAIR OF VENTRICULAR SEPTAL DEFECT IN SMALL CHILDREN

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ABSTRACT

Objective: Significant changes have occurred in the management of pediatric patients with ventricular septal defect such as early surgical repair of the defects. The objective of this study was to investigate the effects of early closure of isolated ventricular septal defects on pulmonary artery pressure.

Materials and Methods: Forty-nine patients undergoing surgery due to ventricular septal defects between January 2001 and January 2004 were enrolled in the study. The patients were classified into two groups in terms of their age at the time of the operation. Those who had been operated on before the age of 2 formed Group 1 (n=24) and those who had been operated on after the age of 2 formed Group 2 (n=25). Preoperative and postoperative (at 1 year) echocardiographic measurements of pulmonary artery pressure values were recorded for all patients.

Results: No statistically significant differences were found between groups 1 and 2 in terms of preoperative pulmonary artery pressure or pulmonary vascular resistance values. However, mean postoperative pulmonary artery pressure values were significantly lower in Group 1 (24.41 ± 0.63 mmHg) when compared with those in Group 2 (37.06 ± 1.53 mmHg) ($p = 0.0001$). No early or late mortality occurred in the groups during a mean follow-up period of 2 years.

Conclusion: Our results suggest that surgery is more beneficial at an earlier age before the onset of pulmonary vascular disease. Surgery at an early age, if possible during infancy, may offer an opportunity to lead a nearly normal life for patients with VSD.

Key Words: Ventricular Septal Defect, Pulmonary Hypertension, Surgical Repair.

KÜÇÜK ÇOCUKLARDA VENTRİKÜLER SEPTAL DEFEKTİN ERKEN CERRAHI ONARIMININ PULMONER ARTER BASINCI ÜZERİNDEKİ ETKİLERİ

ÖZ

Amaç: Ventriküler septal defektli pediatrik hastaların tedavisinde, defektlerin erken cerrahi onarımı gibi önemli değişiklikler olmuştur. Bu çalışmanın amacı, izole ventriküler septal defektlerin erken kapatılmasının pulmoner arter basıncı üzerindeki etkilerinin araştırılmasıdır.

Gereç ve Yöntem: Ocak 2001 - Ocak 2004 arasında ventriküler septal defekt nedeniyle cerrahi tedavi uygulanan 49 hasta çalışmaya dahil edildi. Hastalar ameliyattaki yaşlarına göre iki gruba ayrıldı. İki yaşından önce operasyon edilen hastalar Grup 1 (n=24), 2 yaşından sonra operasyon edilenler Grup 2'yi (n=25) oluşturdu. Preoperatif ve postoperatif (1.yıl) ekokardiyografik pulmoner arter basıncı değerleri tüm hastalar için kaydedildi.

Bulgular: Preoperatif pulmoner arter basıncı ve pulmoner vasküler rezistans değerleri açısından grup 1 ve 2 arasında istatistiksel olarak anlamlı bir fark bulunmadı. Buna karşın, ortalama postoperative pulmoner arter basıncı değerlerinin Grup 1'de ($24,41 \pm 0,63$ mm-Hg) Grup 2 ($37,06 \pm 1,53$ mm-Hg) ile karşılaştırıldığında belirgin olarak düşük olduğu gözlemlendi ($p = 0.0001$). Gruplarda ortalama iki yıllık izlem süresince erken veya geç mortalite oluşmadı.

Sonuç: Sonuçlarımız operasyonun erken yaşta, pulmoner vasküler hastalık başlangıcından önce daha yararlı olduğunu göstermektedir. Erken yaşta, mümkünse infant döneminde cerrahi tedavi VSD'li hastalar için normale yakın bir yaşam sürme şansını sağlayabilmektedir.

Anahtar Kelimeler: Ventriküler Septal Defekt, Pulmoner Hipertansiyon, Cerrahi Onarım.

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INTRODUCTION

The most common congenital heart defect is ventricular septal defect (VSD) and surgical closure of VSDs is one of the most common open heart surgery procedures performed in pediatric patients.¹⁻⁵ It is observed in 25-30% of all children born with a congenital heart defect.⁶ Many VSDs are small and asymptomatic and are likely to close spontaneously.⁷ Unrestricted VSD can cause significant morbidity and mortality in infants due to congestive heart failure and recurrent lung infections.⁸ If left untreated, unrestricted VSD may lead to pulmonary hypertension and pulmonary vascular disease, endocarditis, nutritional defects, and progressive aortic valve regurgitation.^{8,9} Elevated pulmonary vascular resistance in VSD patients with higher left-to-right shunt and eventual development of pulmonary vascular disease are the major restrictive factors limiting life-quality during the course of this condition.¹⁰ Some authors reported that closing large VSDs within the first year of life had favorable effects on infants' haemodynamic and physical development.¹⁰⁻¹³

It has been reported that early VSD closure decreased pulmonary infections and nutritional defects as well as inhibiting the development of right ventricular hypertrophy and arrhythmia.^{8,14}

Patients who underwent VSD closure at early (before 2 years of age) and late (after 2 years of age) stages were compared using echocardiographic evaluations in the preoperative and postoperative period in the present study. Consequently, the impact of age at the time of the operation on postoperative pulmonary artery pressure was assessed comparatively between the groups.

MATERIALS AND METHODS

A retrospective analysis was performed of 49 consecutive patients who had undergone repair of isolated VSD at Gazi University Hospital between January 2001 and January 2004.

The study excluded infants and children with associated cardiac anomalies. The diagnosis of VSD and the decision to operate were exclusively based on a clinical examination, chest radiography, electrocardiography, Doppler echocardiography, and cardiac catheterization. The VSD cases were defined by their adjacent areas as an inlet, trabecular, perimembranous, or outlet defect.

Twenty-four repairs (49%) were performed at or before 2 years of age (Group 1) and 25 repairs (51%) were performed after 2 years of age (Group 2). Table 1 presents the preoperative and operative data for groups 1 and 2.

All patients were operated on by means of cardiopulmonary bypass with aortic and bicaval cannulation and moderate hypothermia (28-30 °C). The VSD was approached through the right atrium and tricuspid valve in all patients. The septal or the anterior

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Table 1: Preoperative data.

	Group 1 (n=24)	Group 2 (n=25)	p Value
Age (months)	11.6 ± 1.4 (3 – 24)	103.2 ± 4.4 (36-144)	p < 0.001
Gender			
Male	17 (70.8%)	18 (72%)	NS
Female	7 (29.2%)	7 (18%)	
PVR	4.47 ± 0.19	4.46±0.17	NS
PAP (mean)	51.12 ± 1.71	55.08 ± 3.22	NS

Table 2: Mean values of pulmonary artery pressure established in the preoperative and postoperative stages for groups 1 and 2.

	Group 1	Group 2	P Value
Preoperative pulmonary artery pressure	51.12 ± 1.71 (38.00 - 65.00)	55.08 ± 3.22 (28.00 - 90.00)	NS
Postoperative pulmonary artery pressure	24.41 ± 0.63 (21.11 - 30.01)	37.06 ± 1.53 (22.33 - 55.84)	P = 0.0001

or leaflet of the tricuspid valve was detached or incised when necessary in order to visualize the VSD. All defects were closed with a Dacron patch with interrupted polypropylene sutures. The patients with elevated pulmonary pressure were kept under deep anesthesia for 24 hours. They were taken off the respiratory support system when elective conditions were established. The patients were evaluated with echocardiography and cardiac catheterization in the preoperative period and with echocardiography in the postoperative period. Preoperative and postoperative (1-year) pulmonary artery pressure (PAP) values were compared using echocardiography.

Postoperative patient follow-up was conducted by the pediatric cardiology and cardiovascular surgery departments in collaboration. All patients were followed up for a mean of 2 years.

Statistical Analysis

All data were assessed using the Wilcoxon rank sum test and Spearman rank correlation test. A p value of under 0.05 was considered statistically significant.

RESULTS

The mean ages at the time of the operation were 11.6 months (range 4-24 months) and 103.2 months (range 36-144 months) in groups 1 and 2, respectively. The most common type of VSD in our patients was perimembranous VSD (32 patients, 65%). The decision for surgery was taken unanimously for all patients with VSD in the councils of pediatric cardiology and cardiovascular surgery. Indications for early surgery were $Q_p/Q_s > 2$, presence of large VSD, heart failure established clinically, or recurrent pulmonary infection.

The mean cardiopulmonary bypass (CPB) times were 84 ± 24 minutes in Group 1 and 81 ± 26 minutes in Group 2 (p>0.05). The mean aortic cross-clamp times were 38 ± 12

and 36 ± 10 minutes in groups 1 and 2, respectively. No statistically significant differences were observed between the groups in terms of

preoperative pulmonary artery pressure or pulmonary artery resistance values determined by echocardiography and cardiac catheterization (p>0.05). As the patients had not undergone cardiac catheterization in the postoperative period, alterations in their postoperative pulmonary artery pressures were monitored echocardiographically (Table 2).

The postoperative decrease in pulmonary artery pressure observed in Group 1 was significantly greater than that in Group 2 (p = 0.0001). No late mortality was observed during the mean follow-up period of 2 years.

DISCUSSION

Significant changes have occurred in the management of patients with VSD, such as early surgical closure of the defects. In spite of this, such patients are still operated on at a later age, with the presence of PVD leading to major postoperative complications in developing countries. Development of PVD remains a major problem for patients with nonrestrictive and nonoperated VSD. Although a number of articles have reported elevated postoperative pulmonary artery pressure in VSD patients operated on at later stages, few studies have been conducted to investigate the association between the age at operation and alterations in postoperative pulmonary pressures.¹⁴⁻¹⁶

Ross-Hesselink et al reported that the most prevalent pathologies leading to death observed in the long-term follow-up of VSD patients were right ventricular hypertrophy and subsequent ventricular arrhythmia. The same study also stated that right ventricular hypertrophy had developed parallel to the elevation in pulmonary vascular resistance.¹⁴



Figure 1: An image during surgery of a patient with a large subaortic ventricular septal defect.

In healthy individuals, high pulmonary vascular resistance at birth recedes to normal values within the first 1-2 weeks.¹⁷ However, this may take longer in nonrestrictive VSD patients and pulmonary vascular resistance may increase even more due to the increase in blood circulation. Increasing resistance may lead to permanent changes in the pulmonary vascular bed¹⁸.

Right ventricular pre-load and after-load increase following the elevation in resistance, resulting in right ventricular hypertrophy. As stated by Ross-Hesselink et al, the level of right ventricular hypertrophy observed in a patient is an indication of that patient's long-term postoperative life quality.¹⁴

When this information is considered, the favorable effects of operating on patients with nonrestrictive VSD in particular at an early age can be better appreciated. Although surgical repair during infancy has favorable effects on babies with VSD, negative effects of open-heart surgery at an early age on mortality have also been presented in the literature.^{19,20}

We maintain that repair of VSDs in patients at an early age will have equally good or in some cases even better postoperative prognosis when compared with those who undergo surgery at a later age, provided suitable conditions are established in the intensive care unit. We are also of the opinion that early operations have very favorable effects on the development of an infant as unresolved and recurrent pulmonary infections can be completely eliminated, as reported in the literature.⁸ Infants who undergo such surgery have been established to rapidly catch up with their peers in terms of weight and height.



Figure 2: Subaortic ventricular septal defect closed by using Dacron patch.

Furthermore, it is evident that those who undergo early VSD surgery will be protected against right ventricular hypertrophy and arrhythmia, leading to late postoperative mortality. The mean pulmonary artery pressure values were observed to be lower in patients undergoing early surgery when compared with those who underwent late surgery in this clinical study.

As a result of advances in diagnostic methods and educational levels of families, it has possible to diagnose VSD at an earlier age, making earlier surgery possible.²¹

Nowadays certain centers are able to offer VSD repair during the infant stage by using only Doppler echocardiography without requiring cardiac catheterization.²² However, Doppler echocardiography cannot establish the pulmonary vascular resistance or Qp / Qs, which are the main indications for VSD surgery. Moreover, certain other cardiac abnormalities (persistent left superior vena cava, aorto-pulmonary collateral artery, etc.) cannot be diagnosed. Therefore, cardiac catheterization becomes essential in most cases. Although cardiac catheterization is still regarded as the gold standard for the preoperative diagnostic approach, advances in noninvasive examinations offer valuable possibilities in the postoperative stage for patient follow-up. Consequently, in this study, postoperative follow-up and PAP measurements were carried out using Doppler echocardiography in patients who did not have postoperative cardiac catheterization due to ethical reasons. Surgical techniques developed for infants, advances in extracorporeal circulation, and improved patient care in the postoperative stage have certainly been of great assistance in carrying out VSD repair at earlier ages.²¹ These advances have no doubt been achieved by the combined efforts of pediatric cardiology, cardiovascular surgery, and intensive care staff. In fact, authors in developed countries do not consider the postoperative stage as a risk factor for surgery at a very early age.^{8,23}

Unrestrictive VSD often leads to complications with the development of PVD. Moderately high PVR results in increased early stage operative mortality and morbidity.¹⁹ Alterations in PVR in the postoperative stage vary greatly. Whi-

le certain investigators^{14,19} demonstrated significantly decreased PVR in the postoperative stage, others^{21,24} reported only slight decreases in PVR. Most of the articles suggesting that PVR rarely decreased in the postoperative stage have either been published 20-25 years ago or have been reported by investigators from developing countries. Most patients enrolled in those studies were undergoing surgery after the age of 2 or 3 years. Early diagnosis is difficult in developing countries as socio-cultural levels and economic resources are low, and many women do not give birth in healthcare centers.

Nowadays, the postoperative stage may be less troublesome for patients with high PVR thanks to the use of certain novel drugs. Beneficial effects of drugs such as prostacyclin, nitric oxide, and sildenafil on patients with high PVR have been reported in the literature.^{21,25,26} The use of these drugs both in the intraoperative and postoperative stage may lead to decreased PVR, chiefly by inducing vasodilatation in the pulmonary vascular bed.

The decrease in pulmonary artery pressure in the postoperative period may not be similar for all patients undergoing VSD repair with similar PVR values according to our clinical observations. Excessive blood flow induced by VSD may not lead to the same alterations in the pulmonary vascular bed in all patients. Thus, we think that certain individual factors may play a role in the development of PVD.

In conclusion, we maintain that carrying out surgery at an earlier age rather than at a later age is more beneficial, particularly for patients with nonrestrictive VSD. Development of pulmonary vascular disease can be restrained, growth as well as physical development at a near-normal level can be ensured, and late stage right ventricular hypertrophy and ventricular arrhythmias can be prevented by early surgery. Therefore, surgery at an early age, if possible during infancy, may offer an opportunity to lead a nearly normal life for patients with VSD.

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