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DEMOGRAPHIC CHARACTERISTICS, HEMODYNAMIC PROPERTIES, TREATMENTS AND OUTCOMES OF PATIENTS WITH ISOLATED VENTRİCULAR SEPTAL DEFECT: A RETROSPECTIVE STUDY

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Abstract

Determine the demographic characteristics, hemodynamic properties, treatments, prognosis and complications of patients diagnosed with isolated ventricular septal defect (VSD). The study was a retrospective study of 268 patients diagnosed with isolated VSD in OMUTF Department of Pediatric Cardiology between January-2010 and December-2018. The cases included in the study were accessed from the database of our hospital. The statistical analysis of data was processed with SPSS v.21 program. 56.3% of the patients were female and 43.7% were male. The most frequent period in which VSDs were diagnosed (50%) was the first 30 days of life. Muscular VSD was the most common type (56.3%). Small VSDs were detected in 83.6%. Membranous defects have a higher tendency to be larger (35.8%). Murmur was detected in 91%. Spontaneous closure was observed in 44.4%, and the rate of spontaneous closure in the first 2 years of life was 58.8%. The spontaneous closure rate of muscular defects was higher. Surgical closure was performed for 5.5%. The need of surgical closure for large membranous defects was higher. Pulmonary hypertension was detected in 2.9% of the patients. VSD is one of the common acyanotic congenital heart diseases. Although it is generally asymptomatic, it can develop to pulmonary hypertension and Eisenmenger syndrome. In case of delayed diagnosis time, the need for medical

or surgical treatment will not be met. The complication and mortality rates of surgical treatment are low. However, we think that it is important to inform families before surgery

Keywords: Ventricular septal defect, congenital heart disease, pediatric cardiology

Özet

İzole ventriküler septal defekt (VSD) tanısı koyulan hastaların demografik özelliklerini, hemodinamik durumlarını, aldıkları tedavileri, prognozları ve gelişen komplikasyonlarını saptamaktır. Ondokuz Mayıs Üniversitesi Tıp Fakültesi (OMÜTF) Çocuk Kardiyoloji Bilim Dalı'na Ocak 2010-Aralık 2018 tarihleri arasında başvuran izole VSD tanısı almış 268 hasta retrospektif olarak incelenmiştir. Vakalara hastanemizin veri tabanından ve ekokardiyografi bilgilerinin kayıtlı olduğu arşiv dokümanlarından ulaşıldı. Verilerin istatistiksel analizi SPSS v.21 programı ile yapılmıştır. Hastaların %56,3'ü kız ve %43,7'si erkektir. En sık tanı koyulan dönem (%50) hayatın ilk 30 günü olmuştur. En sık (%56,3) müsküler tip VSD saptanmıştır. Hastaların %83,6'sında küçük VSD saptanmıştır. Membranöz defektlerin geniş olma oranı daha yüksek (%35,8) bulunmuştur. Hastaların %91'inde üfürüm saptanmıştır. Hastaların %44,4'ünde defektin kendiliğinden kapandığı görülmüştür. İlk 2 yaşta kendiliğinden kapanma oranı %58,8'dir. Müsküler defektlerin kendiliğinden kapanma oranı membranöz tip VSD'lere göre daha yüksektir. Hastaların %5,5'ine cerrahi kapatma uygulanmıştır. Geniş membranöz defektlerde cerrahi tedavi ihtiyacı daha yüksektir. Pulmoner hipertansiyon, hastaların %2,9'unda saptanmıştır. Ventriküler septal defekt sık görülen asiyanotik konjential kalp hastalıklarından birisidir. Genel olarak asemptomatik olsa da pulmoner hipertansiyon ve Eisenmenger sendromuna ilerleyebilir. Bu nedenle hastaların yakın izlemi gerekmektedir. Hastalar zamanında tanı almaz ve takip edilmezlerse medikal veya cerrahi tedavi ihtiyacı karşılanmayacağı için VSD komplikasyonları ortaya çıkabilir ve bir zaman sonunda Eisenmenger sendromu gelişmesiyle cerrahi tedavi şansı kaybedilebilir. Cerrahi tedavinin komplikasyon ve mortalite oranları düşüktür; ancak enfeksiyon gibi kardiyak neden dışındaki komplikasyonlar nedeniyle de mortalite gelişebileceğinden ailelerin cerrahi öncesinde detaylı bir şekilde bilgilendirilmesinin önemli olduğunu düşünmekteyiz.

Anahtar Kelimeler: Ventriküler septal defekt, Konjenital kalp hastalığı, Pediatrik kardiyoloji

1. Introduction

Ventricular septal defect (VSD) is defined as a hole in the interventricular septum, which is the wall separating the right and left ventricles. The interventricular septum (IVS) is a complex structure both embryological and anatomical. Determining the size of the defect, the amount of shunt, and its relationship with other anatomical structures is important for the treatment and prognosis of the disease. It is the most common cardiac malformation seen in childhood. It can occur isolated or accompanied with other congenital heart diseases. Therefore, it is not easy to determine the exact prevalence of VSD. With the increased use of echocardiography and color Doppler, the incidence of VSD has increased. Ventricular septal defect is usually small and asymptomatic and tends to close spontaneously within the first two years of life. Defects that do not close spontaneously can be detected by echocardiography based on their size and hemodynamic characteristics, may require medical treatment, or may need surgical repair. Numerous studies on ventricular septal defect have been conducted both in the past and in present (Anderson et al, 2019). The aim of our study is to determine the demographic characteristics, hemodynamic features, treatments, prognoses, and complications of patients diagnosed with isolated ventricular septal defect who applied to Ondokuz Mayıs University.

2. Material and Methods

The study was conducted to retrospectively analyze 268 patients diagnosed with isolated ventricular septal defect who referred to the Pediatric Cardiology Department of Ondokuz Mayıs University Faculty of Medicine (OMUTF) between January 2010 and December 2018. The cases included in the study were accessed from our hospital's database, notebooks containing echocardiographic information, and archive files. Approval for the study was obtained from the Ethics Committee of Ondokuz Mayıs University on 13.12.2019 with decision number 2019/848. The study included patients diagnosed at OMUTF, aged 0-18 years, with no accompanying other systemic diseases or other cyanotic congenital heart diseases hemodynamic significant. Additionally, patients whose follow-up continued at our outpatient clinic, whose surgeries were performed at our center, who came for regular follow-ups, who applied between January 2010 and December 2018 and who had chromosomal anomalies related to VSD were included in the study.

In our study, the following parameters were examined and compared with the literature: The patients' gender, age, age at diagnosis, presenting complaints, type of VSD, size of the defect, spontaneous closure, gradient over the shunt, other hemodynamically unsignificant cardiac pathologies accompanying at the time of diagnosis, chromosomal anomalies, heart failure, medical treatment, pulmonary hypertension, undergoing surgery, postoperative residuals, blood values (hemoglobin, hematocrit, mean corpuscular volume, mean platelet volume, platelet count), chest radiography, and whether Eisenmenger syndrome developed.

VSD sizes were calculated according to body surface area (Table 1) (Turgeon et al, 2020).

Table 1. Classification of VSD Sizes According to Body Surface Area

Small VSD		**Large VSD**	
< 1 year	≤ 10 mm/m^2	<3 months	≥ 20 mm/m^2
1-6 years	≤ 9 mm/m^2	3 months-6 years	≥ 16 mm/m^2
> 6 years	≤ 5 mm/m^2	> 6 years	≥ 9 mm/m^2

The flow gradient across the ventricular septal defect was grouped as "0-15 mmHg," "15-40 mmHg," and ">40 mmHg." Patients with non-hemodynamically significant cardiac anomalies, including "small ASD, PFO, PDA, PR, PS, MR, AR, AVP, and APCA," were included in the study. Patients whose chest radiographs were not taken appropriately were excluded from the study. Therefore, the evaluated radiographs were those taken in a proper postero-anterior position, at an appropriate distance between the X-ray cassette and the patient, in the correct position at the end of inspiration, with the T3 vertebra's spinous process equidistant from the sternoclavicular joints. The cardiothoracic ratio (CTR) was calculated for patients with appropriately taken radiographs.

For blood values, the patients' hemoglobin (Hb), hematocrit (Hct), mean corpuscular volume (MCV), platelet count, and mean platelet volume (MPV) values measured before and after the operation were recorded and evaluated according to the age-specific ranges of healthy children. Patients who received perioperative erythrocyte suspension (ES) were also included. Values before and after receiving ES were recorded. For the values after receiving ES, measurements taken at least 3 months post-transfusion were included.

The data collected in the research were subjected to statistical analysis using SPSS v.21 software. During data analysis, frequency distribution, percentage calculations, cross-tabulation with chi-square analyses for comparisons between categorical variables, and Mann-Whitney U test for comparing the means of dependent variables with independent variables were conducted, especially for data that did not exhibit a normal distribution.

3. Results and/or Discussion

Our study, which examined patients with isolated ventricular septal defect who referred to the outpatient clinic between January 2010 and December 2018, observed an increase in the number of patients diagnosed with VSD after 2015. Between 2010 and 2015, the number of patients diagnosed was 117 (43.7%), while between 2016 and 2018, this number was 151 (56.4%). The reasons for the increase in the number of diagnosed cases can be attributed to the widespread use of echocardiography and increased screening during the neonatal period. It was observed that 56.3% of the patients were female and 43.7% were male. According to various studies, the female-to-male ratios are similar (Eyileten et al, 2017).

Patients with ventricular septal defect are most diagnosed during the neonatal period and between 1-3 months of age. However, when we look at the ages at diagnosis in our study, we find that 50% of the patients were diagnosed between 0-30 days, 28.4% between 1-6 months, 5.2% between 6-12 months, 6% between 12-24 months, 5.6% between 24 months and 6 years, and 4.9% were diagnosed at age 6 or older. These findings are consistent with the literature (Gersony et al, 2001).

Due to the lack of recorded body weights and heights at the time of diagnosis, meaningful data regarding growth and development retardation could not be obtained. Meaningful data related to the cardiothoracic ratio (CTR) could not be obtained from chest radiographs taken at the time of diagnosis. Chest radiographs began to be uploaded to the system from 2014 onwards. Chest radiographs were not appropriately taken in most patients. Therefore, assessment for cardiomegaly could not be performed.

The most common type of ventricular septal defects seen in the literature is perimembranous (Park et al, 2020). However, muscular defects are more common during the neonatal period. In our study, it was observed that 56.3% of the patients had muscular VSD, 39.6% had membranous VSD, and 4.1% had both muscular and membranous type VSD.

In literature, 72% of patients had small defects, 20.5% had medium-sized defects, and 7.3% had large defects (Turner et al, 1999). In our study, however, it was found that 16.4% of patients had large VSDs, while 83.6% had small ones. We believe that this inconsistency may be due to 50% of our patients being neonates. Consequently, the muscular type is more commonly seen during the neonatal period in our study. Therefore, the proportion of small VSDs was found to be higher in our study as well.

Small VSDs are typically asymptomatic and the diagnosed while detection of a murmur during physical examination. Clinical symptoms of congestive heart failure (CHF) emerge in moderate and large defects (Muralidaran et al, 2019). When examining the complaints reported by patients in our study, it was found that 0.4% experienced decreased feeding, 1.1% had developmental delay, 0.4% felt fatigue, 0.4% had fatigue and developmental delay, 0.4% had hemangioma, 0.4% had hypotension, 3.7% exhibited bruising, 1.1% had a syndromic appearance, 0.4% experienced respiratory distress, 0.4% had tachycardia, 91% presented with a murmur, and 0.4% had both a murmur and bruising.

Ventricular septal defects can spontaneously close at any age. While the closure rate is high within the first two years of life, the rate of spontaneous closure decreases after the age of 10. In our study, when we looked at the time of spontaneous closure of VSD, it was observed that 0.8% closed between 0-30 days, 12.6% between 1-6 months, 26.9% between 6-12 months, 18.5% between 12-24 months, 32.8% between 24 months and 6 years, and 8.4% at age 6 or older. Our findings are consistent with the literature. There is a significant relationship was found between VSD types and spontaneous closure. The rates of spontaneous closure were 57.6% for muscular VSDs, 29.2% for membranous VSDs, and 9.1% for muscular + membranous VSDs. In our study, the rates of spontaneous closure for muscular VSDs were found to be significantly higher, consistent with the literature.

Membranous defects require more surgical intervention than other types. In our study, all 15 patients who underwent surgery had membranous VSDs. Two patients initially had small membranous defects, but they underwent surgery due to enlargement of the VSDs and development of heart failure during follow-up. All patients received anti-congestive therapy before surgery. Patients who do not respond to heart failure treatment require closure of the defect either by catheterization or surgery. If timely intervention is not provided, pulmonary vascular disease and Eisenmenger syndrome may develop (Rao PSM, 2018). Surgery was

performed on 14 patients with congestive heart failure. Percutaneous closure is not performed for VSDs at our center. Among the patients who underwent surgery, congestive heart failure was present in 93.3% of them. In one patient, despite the large defect size, heart failure did not develop. Pulmonary hypertension was present in six patients. Congestive heart failure and pulmonary hypertension improved in all patients during the postoperative period. None of the patients developed Eisenmenger syndrome. Early diagnosis of patients seems crucial for their subsequent management. It has been observed that timely treatment reduces the risk of developing Eisenmenger syndrome or even eliminates it. Patients with large defects or hemodynamically significant defects require close monitoring to detect CHF and complications before they develop. This way, patients can be caught before CHF and its complications develop.

Pulmonary hypertension (PHT) is the most important complication of VSD. There is a significant relationship between the size of the defect and pulmonary hypertension (Noori et al, 2019). In our study, we found 8 patients with pulmonary hypertension. There were no patients with severe pulmonary hypertension. All patients had mild pulmonary hypertension, with 7 of them having membranous and one having muscular VSDs. Pulmonary hypertension regressed in all patients.

Patients with ventricular septal defects have not been extensively studied regarding their blood parameters. In our study, there were 15 patients who underwent surgery. Hemoglobin (Hb), hematocrit (Hct), mean corpuscular volume (MCV), platelet count, and mean platelet volume (MPV) values were examined according to age groups, and no significant difference was observed. All patients received erythrocyte suspension, with one receiving it preoperatively and the rest perioperatively. To investigate the effect of surgical closure of the defect on blood parameters, blood values obtained after at least 3 months following erythrocyte suspension were scanned from the hospital information system, and results from 8 out of 15 operated patients were obtained. There was no significant difference in Hb, Hct, MCV, platelet count, and MPV averages among patients who did not undergo surgery according to VSD size. When comparing Hb values of patients operated between 1-6 months and healthy children, it was found that the Hb value of operated patients was lower. The number of patients undergoing surgery at our center is limited, and blood values have not been regularly monitored. Similarly, blood values were not checked for all patients who did not undergo surgery during routine examinations. Therefore, our findings are limited in terms of evaluation.

After surgical closure of VSD, residual VSD may persist. In our study, residual defects were not found to be hemodynamically significant, and there was no need for further surgical intervention. The authors have noted a high rate of residuals due to their use of TEE (Transesophageal echocardiography) (Schipper et al 2017). In our study, there were 4 patients (26.6%) with residual defects in the postoperative period. Three of them had minimal postoperative residuals persisting, while in one patient, the residual disappeared in the seventh year postoperatively. None of the patients had hemodynamically significant residuals.

4. Conclusion

VSD is one of the common acyanotic heart diseases in childhood. Although generally asymptomatic, it can progress to pulmonary hypertension and Eisenmenger syndrome. Therefore, close monitoring of patients is necessary. If patients do not receive timely diagnosis and follow-up, they may miss the chance for timely and effective medical or surgical closure. The complications and mortality rates of surgical treatment are low; however, mortality can occur as a result of non-cardiac complications such as infection, highlighting the importance of explaining details to families before surgery.

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