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Original Article

The Anomalies of Systemic Venous Connections in Children with Congenital Heart Disease

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ABSTRACT

Anomalous systemic venous return (AVSR) may sometimes present as pathologic entities itself, or associated with other congenital heart defects. The presence of AVSR may require significant changes in surgical technique during the repair of congenital heart defects. The aim of our study was to assess the prevalence of anomalous systemic venous return and to determine the congenital heart anomalies accompanying AVSR in children. This study is based on a retrospective review of the medical records of 175 children who consecutively underwent cardiac catheterisation because that congenital heart disease. The most extensively diagnosed cardiac malformation was VSD (26.8%). ASVR was prevalent in 4.5% of patients. The most commonly diagnosed ASVR was persistent left superior vena cava (3.4%) and a second ASVR type was associated interruption of the inferior vena cava (IVC) with azygos vein continuation (1.1%). A systematic study of systemic venous connections should be performed in all those undergoing cardiac catheterisation as a prelude to open heart repair because it may require significant changes in surgical technique during the repair.

Key words: *Anomalous systemic venous return, congenital heart disease, children*

Doğuştan Kalp Hastalığı Olan Çocuklardaki Sistemik Venöz Dönüş Anomalileri

ÖZET

Anormal sistemik venöz dönüş (AVSR) bazen tek başına patolojik bir durum veya diğer doğumsal kalp hastalıkları ile birlikte görülebilir. AVSR mevcudiyeti doğuştan kalp kusurlarının onarımı sırasında cerrahi teknikte önemli değişiklikler gerektirebilir. Çalışmamızın amacı, doğuştan kalp kusurları olan hastalarda anormal sistemik venöz dönüş sıklığını değerlendirmek ve AVSR'ye eşlik eden konjenital kalp anomalilerini belirlemekti. Bu çalışma doğuştan kalp kusurları nedeniyle kardiyak kateterizasyon uygulanan ardışık 175 çocuğun tıbbi kayıtlarının geriye dönük yorumuna dayanmaktadır. En yaygın kardiyak malformasyon tanısı ventriküler septal defekt idi (26.8%). Anormal sistemik venöz dönüş hastaların % 4.5'inde görüldü. En sık görülen ASVR sol persistan superior vena kava (% 3.4) ve ikinci sıradaki ASVR tipi azygos ven devamlılığı ve kesintili inferior vena kava (% 1.1) idi. AVSR doğuştan kalp kusurlarının onarımı sırasında cerrahi teknikte önemli değişiklikler gerektirebilir bu yüzden açık kalp cerrahisi uygulanması için kardiyak kateterizasyon yapılan tüm hastalara sistemik venöz bağlantı için sistematik bir çalışma yapılmalıdır.

Anahtar kelimeler: *Anormal sistemik venöz dönüş, konjenital kalp hastalığı, çocuk*

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INTRODUCTION

Anomalous systemic venous return (ASVR) is an exceptional form of congenital heart disease in which all of the systemic veins, including the hepatic veins and the coronary sinus, drain abnormally into the heart (1). It is usually anatomically benign, and commonly does not imply haemodynamic consequences. However, ASVR is sometimes associated with atrioventricular canal, common atrium, atrial septal defect (ASD), ventricular septal defect (VSD) and heterotaxia, and it may necessitate important modifications in the surgical technique used during the repair of associated congenital heart defects. Therefore, precise definition of systemic venous connections is important in congenital heart disease to enable proper surgical correction (2). In this study, we present venous anomalies in children with congenital heart disease after catheter angiography over a 3-year period.

MATERIALS AND METHODS

Our study is based on a retrospective review of the medical records of 175 children who consecutively underwent cardiac catheterisation, due to congenital heart disease, at the department of paediatric cardiology in Selcuk University Medical Faculty Hospital, from 2010 to 2013. Transthoracic echocardiography, if necessary, electrocardiography and chest X-ray were performed for all of these patients, prior to cardiac catheterisation and angiography. All echocardiographic examinations were conducted using a commercially available echocardiographic machine (Toshiba, Aplio 50, Japan) equipped with 3.5 and 6.5 MHz transducers. Recordings were performed with patients in the supine or left lateral positions. The innominate vein was evaluated on the frontal plane and, if present, a left superior vena cava was visualised from the same position. The right-sided superior vena cava was visualised by employing a suprasternal notch approach. An enlarged coronary sinus could be demonstrated from a parasternal long axis view. Cardiac catheterisation was performed percutaneously at the right femoral vein, following premedication and local anaesthesia. The innominate vein injection was conducted during cardiac catheterisation, according to the routine practice of the study centre. Data regarding the type of congenital heart disease and persistence of the caval system were listed for each patient following diagnostic procedures, and the Statistical Package for Social Sciences (SPSS for Windows Version 15.00, Chicago, IL, USA) programme was used to assess the results. Distribution of data was analysed using the Kolmogorov-Smirnov test, according to which suitable parametric and non-parametric tests were selected. The parametric data are given as arithmetic means±standard deviation, and non-parametric data are given as medians (minimum–maximum). The Pearson chi-square test was used to compare categorical variables and comparison between groups was determined by Student's t test or Mann–Whitney U test (parametric data and non-parametric data, respectively). In the statistical evaluations, a p value of $p<0.05$ was regarded as significant.

The study protocol was approved by the local ethical committee. Informed consent was obtained from the participants and/or their guardians.

RESULTS

The study group consisted of 175 patients who consecutively underwent cardiac catheterisation at our study centre. Approximately 50.8% (89 of 175) of these patients were male, while 49.2% (86 of 175) were female, and the age range was 1 month-17 years (6.1±4.2 years). In this series, 173 patients had levocardia and two patients had dextrocardia. The most extensively diagnosed cardiac malformation was VSD (n:47, 26.8%) and another common cardiac malformation observed was ASD (n:45, 25.7%). The other cardiac anomalies found were coarctation of the aorta (n:23, 13.1%), patent ductus

***Table 1**. Congenital heart disease in patients with catheter angiography*

Congenital cardiac anomal	% (n)
Ventricular septal defect	26.8 (47)
Atrial septal defect	25.7 (45)
Coarctation of the aorta	13.1 (23)
Patent ductus arteriosus	8.5 (15)
Tetralogy of Fallot	7.4 (13)
Pulmonary valve stenosis	6.8 (12)
Atrioventricular septal defect	2.8 (5)
Aortic valve stenosis	2.2 (4)
Double outlet right ventricle	1.7 (3)
Tricuspid atresia	1.7 (3)
Hypoplastic left heart	(1)
Ebstein's anomaly	(1)
Mitral stenosis	(1)
Transposition of great artery	(1)
Dilated cardiomyopathy	(1)

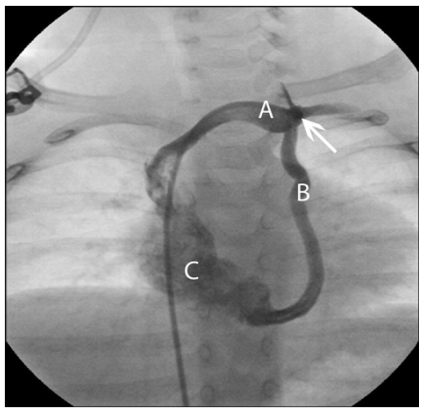

***Figure 1**. Injection in an innominate vein (arrow), connecting the left and right superior vena cava. Posterior-anterior view of the injection showing the catheter in the innominate vein and contrast material in the left superior vena cava and draining into the right atrium. A; innominate vein, B; left superior vena cava, C; right atrium.*

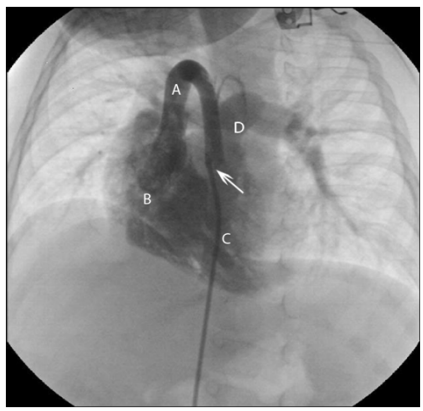

***Figure 2**. Posterior-anterior view shows interruption of the inferior vena cava and the azygous vein that drained into the right atrium in child with ventricular septal defect. The catheter in the azygous vein (arrow). A; azygous vein, B; right atrium C; right ventricle, D; main pulmonary arter.*

arteriosus (n:15, 8.5%), Tetralogy of Fallot (n:13, 7.4%), pulmonary stenosis (n=12, 6.8%), atrioventricular septal defect (n:5, 2.8%), aortic valvular stenosis (n:4, 2.2%), double outlet right ventricle (n:3, 1.7%), tricuspid atresia (n:3, 1.7%), hypoplastic left heart syndrome (n:1), Ebstein's anomaly(n=1), mitral stenosis (n:1), transposition of great arteries (n:1) and dilated cardiomyopathy (n:1). Table 1 demonstrates the congenital heart anomalies diagnosed within our study population. Anomalous systemic venous return was prevalent in 4.5% (8 of 175) of patients with congenital heart disease. The most commonly diagnosed ASVR was persistent left superior vena cava (PLSVC; n:6, 3.4%) (Figure 1) and a second ASVR was associated interruption of the inferior vena cava (IVC) with azygos vein continuation (n:2, 1.1%), as shown in Figure 2. The detected venous return and accompanying congenital heart anomalies are shown in Table 2.

DISCUSSION

Normal systemic venous return consists of the connection of the superior vena cava (SVC) and IVC, as well as the coronary sinus, to the right atrium on the right side of the interatrial septum. The development of this venous system begins from the symmetric and paired cardinal and subcardinal veins in the 5-week-old embryo (3). Countless abnormalities of this system are com-

***Table 2**. Anomalous systemic venous return in relation with congenital cardiac anomalies*

No	Age	Sex	Anomalous systemic venous return	Congenital cardiac anomaly
7	8 months	Male	Persistent left superior vena cava	Ventricular septal defect
24	3 months	Famele	Persistent left superior vena cava	Atrioventricular septal defect
31	11 months	Male	Persistent left superior vena cava	Tetralogy of Fallot
45	8 months	Male	The associated interruption of the inferior vena cava with azygos vein continuation	Atrial septal defect
51	13 months	Famele	Persistent left superior vena cava	Tetralogy of Fallot
68	10 months	Famele	Persistent left superior vena cava	Tetralogy of Fallot
74	2 years	Famele	The associated interruption of the inferior vena cava with azygos vein continuation	Double outlet right ventricle
81	5 years	Male	Persistent left superior vena cava	Ventricular septal defect

monly associated with other cardiac malformations, and systemic venous return manifests as a wide spectrum of structural abnormalities. These malformations are relatively rare, occurring in approximately 0.3% to 0.4% of the general population,4 but, are more prevalent in congenital heart defects, occurring in 2.8% to 4.8% of operative cases (1). Multiple classification diagrams have been proposed for the characterisation and organisation of systemic venous anomalies. However, for the surgeon, the site of abnormal systemic venous cardiac connection has implications for operative intervention, such that the chamber receiving abnormal connections may form the basis of a classification. The Society of Thoracic Surgeons Congenital Heart Surgery Database committee suggested a hierarchical scheme for classification, again based on the IVC, the SVC and the hepatic veins (2). Other physicians have used a systematic approach, based on embryological considerations, emphasising abnormally persisting segments (1).

Persistent left superior vena cava (PLSVC) is a common congenital malformation of the thoracic venous system, with a prevalence estimated at approximately 0.3% in individuals with a normal heart and 4.5% in individuals with congenital heart disease (5). It is generally asymptomatic and is detected when cardiovascular imaging is performed for unrelated reasons. The well-developed azygos or hemiazygos vein is present and connects with some portion of the infrarenal IVC. Azygos continuation of the IVC on the left or right may occur in either situs solitus or left isomerism,(6) but does not necessarily imply absence of the suprarenal inferior cava, which may coexist in patients with situs solitus, although rarely. The bridging innominate vein connects the bilateral SVC, and, when present, runs in the anterior mediastinum, immediately in front of the origin of the brachiocephalic branches of the aorta. Partial anomalous hepatic venous connection is such that some, but not all, of the hepatic veins connect directly to the heart in an abnormal fashion, instead of to the suprarenal inferior cava or to the inferior cavoatrial junction. Total anomalous hepatic venous connection is such that all the hepatic veins connect directly to the heart, via one or more hepatic veins. This may be to the left or the right sided atrium, or to both, and the hepatic veins connect separately to the heart if the suprarenal inferior cava is present. This venous connection was present in all patients with left isomerism (6). The majority of systemic venous anomalies are accounted for by a few specific abnormalities. In one study, the overall incidence of systemic venous return was 3.8%, and 48% of these anomalies were represented by bilateral SVC, while 28% were represented by interrupted IVC (1). In our catheter angiography series, the prevalence of ASVR was 4.5%, and PLSVC was the most common abnormality of systemic venous return. Huhta et al. (7) found ASVR in 53 patients (6%), right SVC in 792/792 patients (100%); left SVC in 46/48 patients (96%); bilateral SVC in 38/40 patients (95%); bridging innominate vein with bilateral SVC in 13/18 patients (72%); connection of superior caval segment to heart (coronary sinus or either atrium) (100%); absence of suprarenal IVC in 23/23 patients (100%); azygos continuation of the IVC in 31/33 patients (91%) and total anomalous hepatic venous connection (invariably associated with left isomerism) in 23/23 patients (100%). In another study, (8) Three hundred and sixty-nine consecutive foetuses were diagnosed as having cardiac malformation, including 27 (approximately 7.3%) that had an ASVR. According to this study, the most common forms of ASVR were PLSVC (n:15, 56%) and interrupted IVC (n:6, 22%). The remaining types of ASVR were PLSVC without a right SVC (n:4, 15%), bilateral agenesis of the SVC, with cephalic venous drainage towards the IVC through the azygos system (n:1) and total ASVR to the coronary sinus (n:1).

Bilateral SVC was reported in 0.3% of the general population in an autopsy series, while SVC has been reported in 3-5% of patients with congenital heart disease (9). Its draining into the right atrium, via the coronary sinus, is one of the more common systemic venous anomalies, and is frequently incidentally. If an abnormality is isolated, this finding is usually not clinically significant; however, it is frequently associated with an atrial septal defect or other intracardiac malformations (10). Although a lot of single case reports exist, studies showing incidence of systemic venous return are rare in the literature. Left SVC was observed in 6/175 (3.4%) of our patients. Azygos and/or hemiazygos continuation of the IVC has been reported to occur in 0.6% patients with congenital heart disease. It is important to define because inadvertent ligation of the azygos vein during surgical procedures must be completely avoided in such situations, and ligation has been known to cause death (9). In our series, infrahepatic interruption of the IVC with azygos or hemiazygos continuation was observed in 2/175 (1.1%) patients with congenital heart disease.

In conclusion, ASVR is a complex and varied group of

lesions that are not uncommon in patients undergoing congenital heart surgery. Furthermore, such anomalies often dictate significant modifications in the conduct and timing of cardiac surgical procedures. The angiographic definition of systemic venous connections may lead to a small increase in the amount of fluoroscopy time during the catheterisation procedure, and the information that is gained is very useful. We believe that, patient's clinical condition permitting, a systematic study of systemic venous connections should be performed in all those undergoing cardiac catheterisation as a prelude to open heart repair.

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