Problems of the neonates with congenital heart disease requiring early interventions: a regional report

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Abstract

Aim: In this study, it was aimed to determine the problems of the neonates who were diagnosed with congenital heart disease requiring early intervention in our hospital.

Material and Methods: The files of the newborn babies with congenital heart disease requiring early intervention who were admitted to the neonatal intensive care unit of our hospital between January 2011 and January 2013 were evaluated retrospectively. In all cases, echocardiography and "Score for Neonatal Acute Physiology-II" (SNAP-II) scoring were performed within the first day of admission. The data were interpreted using Number Cruncher Statistical System 2007 software. The statistical significance was set at $p < 0.05$.

Results: A total of 83 babies were included in the study. Forty six of the patients were male (55%), and 37 (45%) were female. Sixty eight percent of the patients were referred from the neighboring provinces and 32% were transferred from the centers within the city. The age range was between 0 and 28 (5.6 ± 6.4 day) days. The SNAP-II scores upon admission ranged between 0 and 90 (mean: 20 ± 20.3). None of the patients was diagnosed prenatally. The most common diagnoses included transposition of the great arteries (33.7%) and pulmonary atresia (19.3%). Nineteen (22%) patients were lost in the neonatal intensive care unit. There was a significant relationship between the mortality and the SNAP-II scores ($p = 0.0001$) and use of vasopressors ($p = 0.004$). The diagnosis, gender, use of alprostadil and age were not related to mortality. Three patients were discharged following planning of elective surgery and 60 patients were referred to a tertiary center by air ambulance.

Conclusions: The results of our study indicated that prenatal diagnosis could not be made in neonates with congenital heart disease requiring intervention in our region. The mortality rates of these patients were related to the severity of the clinical status at presentation rather than to the age, gender and type of congenital heart disease. The mortality was much higher in the patients who developed circulatory failure. Most of the patients who survived were sent by air ambulance to the centers where the intervention was to be performed. (Turk Pediatri Ars 2015; 50: 158-62)

Keywords: Congenital heart disease requiring intervention, prenatal diagnosis, transport, newborn

Introduction

Congenital heart diseases (CHD) are observed with a frequency of 0.3-1.2% (1). Congenital heart diseases may involve simple defects or severe pictures which require early diagnosis and intervention for survival (2, 3). According to the data of the European Congenital Heart Diseases Committee, it is predicted that CHD requiring early intervention will develop in 0.4% of newborn babies (2). Intervention is needed in the neonatal period in 15-20% of the subjects with congenital heart disease (4, 5). Studies have proposed that identification of CHD in the prenatal period will facilitate

performance of necessary interventions timely and under appropriate conditions and thus reduce the rate of neonatal mortality due to CHD from 2-3/1 000 to 0.6-0.8/1 000 (2, 3). Acidosis has been observed to develop with a lower rate and lactate levels have been observed to be lower in patients diagnosed in the prenatal period and it has been reported that this might have an impact on hospitalization period and long-term neurological development (3).

In this study, it was aimed to determine the demographic data, SNAP-II scores, diagnoses, duration of hospitalization in the intensive care unit, therapies ad-

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ministered and mortality rates in the newborns who were diagnosed with CHD and who required intervention in our unit, to examine the ways of transportation of these subjects to the centers where the interventions would be performed, to draw attention to present negativities and to offer solutions.

Material and Methods

In this study, 83 newborns who were hospitalized in Diyarbakır Children's Hospital Neonatal Intensive Care Unit (NICU) between January 2011 and January 2013 and diagnosed with CHD requiring intervention were evaluated retrospectively. The subjects were examined in terms of residence city, age, gender, diagnosis, duration of hospitalization in the unit, SNAP-II score, treatment administered, mortality rate and way of referral. Echocardiogram was performed in all patients by a pediatric cardiologist using Philips HDX 11 or General Electric Vivid 4 device. Written and verbal informed consent was obtained from the families of all patients.

Statistical analysis

In this study, the statistical analyses were performed using NCSS (Number Cruncher Statistical System) 2007 Statistical Software (Utah, USA) package program. In assessment of the data, Kruskal Wallis test, Mann-Whitney U test and chi-square test were used in comparison of the variables which did not show a normal distribution in addition to descriptive statistics (mean, standard deviation). Logistic regression analysis was used to determine the factors which affected the loss and the area under the ROC curve was calculated to determine the effect of Snap II score in determining the loss. A p value of <0.05 was considered significant.

Results

Forty six (55%) of the subjects were male and 37 (45%) were female. Twenty seven subjects were referred to our unit from Diyarbakır and 56 were referred from the neighboring provinces (Table 1). None of the subjects were diagnosed in the prenatal period. The mean age of presentation was 5.6 ± 6.4 days (0-28 days). The mean period of hospitalization in the unit was 4.3 ± 6.6 days (0-41 days). The mean SNAP-II score at presentation was found to be $20 \pm 20,3$ (0-90). Common diagnoses included transposition of the great arteries (33.7%) and pulmonary atresia (19.3%) (Table 2). Prostaglandin was given to 46 (55%) of the subjects who had ductus dependent systemic or pulmonary circulation, vasopressor was given to 25 subjects (30%) who had circulatory

Table 1. Distribution of the patients by gender and province of origin

		n	%
Gender	Male	47	56.6
	Female	36	43.3
Province	Diyarbakır	27	32.5
	Mardin	17	20.4
	Batman	12	14.4
	Şırnak	10	12.0
	Siirt	3	3.6
	Bitlis	3	3.6
	Bingöl	3	3.6
	Muş	3	3.6
	Hakkari	3	3.6
	Adıyaman	1	1.2
	Şanlıurfa	1	1.2

Table 2. Distribution of the patients by diagnosis

		n	%
Diagnosis	Transposition of the great arteries	28	33.7
	Pulmonary atresia	16	19.3
	Aortic coarctation	10	12.0
	Single ventricle	6	7.2
	Hypoplastic left heart syndrome	6	7.2
	Tricuspid atresia	5	6.0
	Pulmonary stenosis	3	3.6
	Truncus arteriosus	2	2.4
	Aortic arch anomaly	2	2.4
	Cardiomyopathy	1	1.2
	Taussig Bing anomaly	2	2.4
	Valvular aortic stenosis	1	1.2
	AV complete block	1	1.2
	Total	83	100.0

disorder and vasopressor support plus prostaglandin treatment was given to 12 subjects (14%). Nineteen (22%) patients were lost in our unit. The mortality rate was 26% in the subjects who received only prostaglandin, 52% in the subjects who received only vasopressor and 57% in the subjects who received both prostaglandin and vasopressor. The subjects who were lost had the following diagnoses: transposition of the great arteries (seven subjects), pulmonary atresia (three subjects), single ventricle (three subjects), hypoplastic left heart syndrome (two subjects), tricuspid atresia (one subject), aortic arch anomaly (one subject), Taussig-Bing anomaly (one subject) and cardiomyopathy (one subject). When the subjects who were referred or discharged

Table 3. Comparison of the subjects who were referred or discharged and the subjects who were lost

		Referral+discharge group n:64		Subjects who were lost n:19		p*
Age (days)		6.41±6.8		2.95±4.09		0.062
Gender	Male	38	59.38%	9	47.37%	0.354
	Female	26	40.63%	10	52.63%	
Diagnosis	Group with increased pulmonary blood flow ^a	23	35.94%	7	36.84%	0.998
	Group with decreased pulmonary blood flow ^b	23	35.94%	7	36.84%	
	Group with decreased systemic blood flow ^c	14	21.88%	4	21.05%	
	Other	4	6.25%	1	5.26%	
Weight at presentation (g)		3097.11±517.81		2982.37±611.2		0.186
Duration of hospitalization (days)		3.64±4.08		6.68±11.64		0.842
Prostaglandin		35	54.69%	11	57.89%	0.805
Vasopressor		12	18.75%	13	68.42%	0.0001
SNAP II		13.28±12.59		42.68±24.96		0.0001

*Mann Whitney U test-Chi-square test

^aGroup with increased pulmonary blood flow (Truncus arteriosus, transposition of the great arteries)^bGroups with decreased pulmonary blood flow (Pulmonary stenosis-atresia, tricuspid atresia, Double outlet right ventricle, Single ventricle)^cGroup with decreased systemic blood flow (Aortic coarctation, Aortic stenosis, Aortic arch anomaly, Hypoplastic left heart)**Table 4. The association of SNAP II score and use of vasopressor with mortality-logistic regression analysis**

	B	S.E.	p	OR	%95 CI	
				(Odd Ratio)	Lower	Upper
Snap II	0.08	0.02	0.0001	1.08	1.04	1.13
Vasopressor	-2.11	0.74	0.004	0.12	0.03	0.52
Constant	-1.93	0.67	0.004	0.15		

Table 5. Distribution of the patients by provinces they were referred to

		n	%
Center of referral	Ankara	20	33.3
	İstanbul	19	31.6
	Malatya	12	20.0
	Adana	5	8.3
	Kayseri	1	1.6
	Erzurum	1	1.6
	Kocaeli	1	1.6
	Gaziantep	1	1.6
	Toplam	60	100

and the subjects who were lost were compared, no statistical difference was found in terms of mean ages, gender distributions, body weights at presentation, distributions of diagnosis, periods of hospitalization and use of prostaglandin ($p>0.05$). On the other hand, use of vasopressor and SNAP II scores were found to be statis-

tically significantly higher in the subjects who were lost ($p=0.0001$) (Table 3). Logistic regression analysis was performed with SNAP II score and use of vasopressor to determine the factors having an impact on the mortality. Increased SNAP II score ($p=0.0001$) and use of vasopressor ($p=0.004$) were determined to be statistically efficient factors (Table 4). The area under the ROC curve was calculated to determine the strength of SNAP II score in predicting the risk of loss and (AUC) was found to be 0.871. The prediction point was found to be >21 for a sensitivity of 78.95, for a specificity of 79.69, for a positive predictive value of 53.6 and for a negative predictive value of 92.7. The likelihood ratio calculated for this point was 3.89 (Figure 1).

Sixty subjects were referred to larger centers with air ambulance and three subjects were discharged by planning elective operation (Table 5).

Discussion

Since our unit is not an obstetrics clinic, all our patients were referred from other centers. Since there is no cardiovascular surgery center in our hospital and region, patients with CHD who need intervention are referred to other centers. The patients are transported by way of air ambulance to the cities outside Malatya and by way of ambulance helicopter belonging to Diyarbakır Health Department to Malatya. In the literature, the cost of transportation has been reported to be 6-7 dollars per mile by plane and 7-24 dollars per mile by helicopter. The lowest cost per patient has been reported

Figure 1. Strength of SNAP II score to predict mortality risk

to be 2811 dollars (6, 7). Referral in the prenatal period is recommended in terms of both the patient's health and cost efficiency (8). As far as we know, there is no cost efficiency study related with referral of newborns in our country.

None of the patients included in our study was diagnosed prenatally. They were diagnosed in our unit. This suggests that patients in our region are underdiagnosed in the prenatal period and lose the chance of referral before delivery. Larger studies related with this issue are needed and adjustments which would increase the possibility of making a diagnosis in the prenatal period in this region are needed, because this situation may affect the morbidity and mortality rates of patients.

SNAP-II scoring is a scoring method developed by Richardson et al. (9) to predict the mortality risk in term newborns at the time of admission. In our study, a relationship was found between the SNAP-II score and mortality rates, but no relationship was found between the SNAP-II score and diagnosis, gender and weight at presentation. In patients with a SNAP-II score of >21 , the mortality risk was found to be 3,89-fold higher compared to the patients with a SNAP-II score of <21 . This suggests that our patients are lost in relation with other factors which might affect their general status at admission rather than the type of CHD.

The mortality rate was found to be 26% in the patients who had ductus dependent circulation and received only prostaglandin, 52% in the patients who received vasopressor medication because of circulatory failure and 57% in the patients who had ductus dependent circulation and received vasopressor treatment together

with prostaglandin because of circulatory failure. In the logistic regression analysis, use of vasopressor was found to be related to mortality. These findings suggest that referral of newborns with CHD requiring intervention to intervention centers before circulatory failure develops will decrease mortality rates to a significant extent.

The most common CHD requiring intervention in the newborn period is transposition of the great arteries (10). In our study, transposition of the great arteries was also in the first order in terms of disease distribution and it was not observed that a certain disease was intensified in a certain province. The outcomes of the patients referred for intervention were not evaluated, because this was outside the scope of this study.

According to the results of our study, it was found that newborns with CHD requiring intervention were not diagnosed in the prenatal period in our region, the mortality rates of these patients were related with the clinical status at presentation rather than age and type of CHD, the mortality rates were much higher in the ones with circulatory failure and the majority of the surviving patients were transported to the centers where intervention would be performed by way of air ambulance.

We think that centers where interventional procedures and cardiovascular operations for CHD can be performed, should be established as soon as possible in our country in provinces which are regional centers as recommended by the European Congenital Heart Diseases Committee to solve these problems and these patients should be diagnosed in the prenatal period by way of fetal echocardiogram and referred to the related centers before delivery.

Ethics Committee Approval: Ethics committee approval was not received due to the retrospective nature of the study.

Informed Consent: Written and oral informed consent was obtained from the parent of the patient.

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