Pediatric Emergency Room Presentation of Congenital Heart Disease

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ABSTRACT

Background and Objectives: Only a few studies have specifically investigated the reasons for emergency room (ER) visits in patients with congenital heart disease (CHD). The aim of this study was to identify the major reasons for ER presentation among patients with CHD that were acutely and seriously ill at a tertiary medical center in Korea. Subjects and Methods: All 368 admissions of patients with CHD via the ER from 2003 to 2008 were enrolled. We conducted a retrospective study with review of the medical records. Results: Eighty two patients were newly diagnosed as having CHD. Their major presentations were symptoms of heart failure (41.5%), murmur (31.7%), and cyanosis (18.3%). There were 286 visits that were cases with known CHD. Their major presentations were respiratory tract infection (24.1%, 2.7 ± 4.1 years of age), dysrhythmia (16.4%, 16.7 ± 9.5 years), symptoms of heart failure (14.3%, 7.6 ± 9.4 years), aggravated cyanosis (5.6%, 0.8 ± 1.4 years), protein-losing enteropathy (4.9%), hemoptysis (4.5%), drug side effects (4.1%), and infective endocarditis (3.0%). There were significant correlations between the age distributions and major modes of presentation. Surgical treatments were required within 1 month in 38%, and 2.7% of all patients died during hospitalization. The patient group with respiratory infections and CHD showed the highest mortality (5.8%). Atrial flutter was the most frequent arrhythmia (70.2%) and 70% of these patients were post-Fontan surgery condition. The causes of heart failure in the patients with previous surgical repair were: pulmonary hypertension, myocardial dysfunction, valve regurgitation, and uncorrected lesions. Conclusion: Improved understanding of the common problems in the ER can help prepare clinicians to manage patients that present with CHD. (Korean Circ J 2010;40:36-41)

KEY WORDS: Congenital heart; Emergencies.

Introduction

Acutely compromised patients with congenital heart disease (CHD) can be very complicated for the physician to manage. Patients with CHD do not present with only organ-specific symptoms, but also have generalized symptoms. Making the correct decisions quickly in the ER to manage these patients can be extremely difficult.

There are many prior studies describing the pathophysiology, clinical manifestations and management of patients with CHD. However, only a few studies have specifically addressed the reasons for emergency room (ER) visits among patients with CHD. Recently, the pattern of presentations of CHD in the ER has changed because of additional information from antenatal diagnosis, a more organized inter-hospital referral system, early repair of CHD, and the increased population of adults with CHD.

In this study we evaluated the patterns of clinical symptoms among patients with CHD presenting to the ER at a tertiary medical center in Korea.

Subjects and Methods

We reviewed the medical records of patients with CHD that were admitted to the department of pediatric car-
diology and pediatric thoracic surgery via the pediatric emergency room (PER) of Seoul National University Hospital (SNUH) from May 2003 to April 2008. We included patients with CHD if their symptoms were suspected to be associated with their underlying CHD, previous cardiac surgery or cardiac medications. We excluded patients with no direct relationship between the major problem identified at the PER and their underlying CHD.

The collected data included gender, date of the ER visit, age at the ER visit, presence of a pre-existing diagnosis, presence of a previous surgery, chief complaint, duration of hospitalization, diagnosis, surgery during hospitalization, interval from admission to surgery, operation record, and the patient condition at discharge.

We divided all data into two major groups: Group 1 was ER visits among newly diagnosed patients with CHD, and Group 2 was ER visits among patients with a pre-existing diagnosis of CHD. Statistical analysis was performed using the Student’s t-test for continuous variables and the χ² test for nominal variables (Statistical Package for the Social Sciences (SPSS), version 12.0 (SPSS Inc., Chicago, IL, USA)). A p of <0.05 was considered statistically significant.

Results

There were 368 ER visits among 256 patients with CHD during the study period. During the same period, the number of patients that were admitted via the PER were 8,902. Therefore, 4.1% of all admissions via the PER were for patients with CHD that required hospitalization. Patients had a median age of 1.5 years, with a range of 1 day to 31 years. There were 188 male patients and 180 female patients. The mean duration of hospitalization was 20.0±28.2 days. Ninety five patients (25.8%) underwent cardiovascular surgery within 1 week of hospitalization and 139 patients (37.8%) within 4 weeks of hospitalization. Ten out of all of the patients (2.7%) died within 4 weeks of hospitalization. One of these patients was newly diagnosed with CHD and the other nine patients had a pre-existing diagnosis of CHD (Table 1).

We divided the study population into two major groups—newly diagnosed patients with CHD (Group 1) and pre-existing CHD patients (Group 2). The mean age of the patients in Group 1 was younger than in Group 2, and this difference was statistically significant (3.5±13.7 months vs. 8.6±9.1 years; p<0.001). The proportion of patients that required cardiovascular surgery within one to four weeks after hospitalization was higher in Group 1 than in Group 2 (70.7% vs. 12.9% and 74.3% vs. 20.2%; p<0.001). However, there was no significant difference in the duration of hospitalization or mortality within four weeks of hospitalization between the two groups (Table 2).

Characteristics of the newly diagnosed patients with congenital heart disease (Group 1)

Group 1 (the newly diagnosed patient group) included 82 PER visits where a new diagnosis of CHD was made as a result of the ER visit or subsequent hospitalization. Fifty-five patients among them (79.3%) were referred from other hospitals. The mean patient age was 3.5±13.7 months (median 1.0 month; range 1 day to 4 years); the mean duration of hospitalization was 24.2±31.8 days (median 19 days; range 3 to 121 days).

Table 1. Characteristics of ten patients that died during hospitalization within four weeks after admission via the ER

| Case | Age/SEX | Underlying CHD | Surgery before ER visit | Mode of ER presentation | Surgery after admission | Cause of death |
|------|---------|----------------|-------------------------|-------------------------|------------------------|----------------|
| 1    | 6 w/M   | TGA, ASD       | -                       | Cyanosis                | PAB & BTS              | LV dysfunction  |
| 2    | 23 m o/M| TGA, ASD       | BCPS, AVVR              | Pneumonia               | -                      | Sepsis, respiratory failure |
| 3    | 6 mo/M  | CoA, VSD       | VSD closure, coarctoplasty | Pneumonia, pulmonary congestion | -                      | Progressive multiple PV stenosis, respiratory failure |
| 4    | 5 yr/M  | CoA, VSD       | Fontan operation         | Pneumonia               | -                      | Septic shock, hepatic failure |
| 5    | 18 yr/M | TOF            | Total correction         | Pneumonia               | -                      | Multi-organ failure, massive hemoptysis |
| 6    | 7 yr/M  | Congenital PV stenosis | PV widening      | Heart failure           | -                      | Sudden cardiac arrest, pulmonary hypertension |
| 7    | 15 yr/M | TOF with PA    | Rastelli operation       | Heart failure           | -                      | LV failure, ventricular tachycardia |
| 8    | 11 yr/F | CoA, RI, Kartagener’s syndrome | Fontan operation | Hemoptyis              | -                      | Hemoptyis |
| 9    | 11 yr/F | ECD, Eisenmenger syndrome | PA, TAPVR, ECD, RI | Hemoptyis              | -                      | Hemoptyis |
| 10   | 5 yr/F  | TAPVR, ECD, RI | TAPVR repair, mBTS      | Respiratory difficulty  | -                      | Upper airway problem |

CHD: congenital heart disease, ER: emergency room, TGA: transposition of great arteries, ASD: atrial septal defect, PAB: pulmonary arterial banding, BTS: Blalock-Taussig shunt, LV: left ventricle, 6SV: functional single ventricle, RI: right isomerism, BCPS: bidirectional cavopulmonary shunt, AVVR: atrioventricular valve replacement, CoA: coarctation of the aorta, VSD: ventricular septal defect, PV: pulmonary vein, TOF: Tetralogy of Fallot, PA: pulmonary atresia, ECD: endocardial cushion defect, TAPVR: total anomalous pulmonary venous return, mBTS: modified Blalock-Taussig shunt
Thirty-four patients (41.5%) presented with signs of heart failure such as tachypnea, cardiomegaly, cough or respiratory difficulty without any evidence of respiratory tract infection. Twenty-six patients (31.7%) visited the PER for the presence of a murmur. Fifteen patients (18.3%) had cyanosis and 6 (7.3%) had symptoms of a respiratory tract infection. One patient was transferred to the PER for evaluation of other anomalies and a murmur (Fig. 1A).

Their underlying cardiac lesions included a ventricular septal defect (VSD, 22 cases; 26.8%), aortic lesions such as coarctation of aorta (CoA) or interrupted aortic arch (IAA, with or without VSD) (18 cases; 22.0%), tetralogy of Fallot (TOF, 13 cases; 15.9%), transposition of the great arteries (TGA, 11 cases; 13.4%), a complex univentricular heart (5 cases; 6.1%), total anomalous pulmonary venous return (TAPVR, 3 cases; 3.7%), and miscellaneous lesions (endocardial cushion defect (ECD), patent ductus arteriosus (PDA), atrial septal defect (ASD), pulmonary stenosis (PS)). There were 48 cases (58.5%) with cyanotic congenital heart lesions such as TOF, TGA, a single ventricle, TAPVR, and 34 cases (41.5%) of acyanotic congenital heart lesions such as VSD, aortic lesion, ECD, PDA, ASD, and PS.

Among all of the Group 1 patients, 19 (23.2%) had PDA dependent CHD requiring prostaglandin E1 (PGE1) infusion. All 19 patients visited our PER via referral from other hospitals and PGE1 was started before transfer to our hospital only in 8 (47%) out of 17 patients, where administration of PGE1 prior to transfer could be confirmed by the information in the medical records. Their mean age was 3.2 ± 1.0 years, and 9 of them had CoA/IAA, 5 had TGA, 2 had pulmonary atresia (one with intact ventricular septum and the other with single ventricle), 2 had TOF, and 1 patient had critical PS. The mean interval to operation was 4.7 ± 5.3 days, and none of these patients died during the hospitalization.

Among Group 1 patients, there was one mortality (1.2%) in a 6 week-old male patient with complete TGA with an intact ventricular septum. Because left ventricular thinning had significantly progressed [squeezed left ventricle (LV) with posterior wall thickness of 2.6 mm during diastole], he underwent pulmonary arterial banding and a modified Blalock-Taussig shunt operation for training of the LV. Left ventricular dysfunction developed and he died one day after surgery.

**Characteristics of the patients with known congenital heart disease (Group 2)**

Group 2 (patients known to have CHD) included 286 PER visits among 228 patients. Their mean age was 8.6 ± 9.1 years (median 3.8 years; range 3 to 31.3 years) and the mean duration of hospitalization was 19.4 ± 9.0 days (median 10 days; range 1 to 298 days).

In group 2, the patients visited the PER because of respiratory tract infection (69 visits; 24.1%), dysrhythmia (47 visits; 16.4%), signs of heart failure (42 visits; 14.7%), aggravated cyanosis (15 visits; 5.2%), aggravated symptoms of a protein-losing enteropathy (14 visits; 4.9%),
hemoptyis (13 visits; 4.5%), drug-related complications due to warfarin or digoxin (12 visits; 4.2%), infective endocarditis (9 visits; 3.2%), chest pain (5 visits; 1.8%), and other (60 visits; 21.0%; non-specific fever, shunt obstruction on echocardiogram, and Kawasaki disease) (Fig. 1B).

The age distribution of the four major clinical reasons for ER presentation (respiratory tract infection, dysrhythmia, signs of heart failure, and aggravated cyanosis) revealed significant differences. The patients in the dysrhythmia subgroup had the oldest age distribution (16.7 ± 9.5 years) among the major four subgroups, and this difference was statistically significant (p<0.001). The subgroups with respiratory tract infection (2.7 ± 4.1 years) or cyanosis (0.8 ± 1.4 years) were younger than those with heart failure symptoms (7.6 ± 9.4 years) and dysrhythmia (p<0.001). However, the age distribution among the subgroup with respiratory tract infection and the cyanosis subgroup was not significantly different (p=0.398) (Fig. 2).

Thirty-five patients (15.4%) in group 2 repeatedly visited the PER 93 times with the same problems. The reasons for the repeated PER visits were as follows: respiratory tract infection associated with aggravation of heart failure or hypoxemia (13 patients, 32 visits), dysrhythmia (6 patients, 21 visits), and heart failure (8 patients, 18 visits).

In the subgroup of respiratory tract infection, the mean age of the patients was 2.7 ± 4.1 years and the median age was 1.1 years with range of 27 days to 21.6 years. Their mean duration of hospitalization was 17.6 ± 36.3 days and there were four deaths in this group (5.8%). The four deaths included three patients with pneumonia and one with pneumonia-associated sepsis. The underlying congenital cardiac lesions with respiratory tract infection were TOF (20 cases; 29.0%), CoA (9 cases; 13.0%), VSD, ECD (7 cases in each; 10.1%), double outlet of right ventricle (DORV), PS (6 cases in each; 8.7%), single ventricle or post-Fontan procedure (5 cases; 7.2%), and other (e.g., truncus arteriosus, IAA, and ASD). In 54 cases (78.3%), patients had already undergone previous cardiac surgery before the PER visits and in 15 cases (21.7%), the patients had no history of previous cardiac surgery.

In the subgroup with dysrhythmia, the mean age was 16.7 ± 9.5 years (range 25 days to 30.2 years) with the median age of 17.7 years. The underlying cardiac lesions included a single ventricle or post-Fontan procedure (24 cases; 51.1%), TOF (5 cases; 10.6%), TGA, DORV, ASD (4 cases in each; 8.5%), VSD, ECD (2 cases; 4.3%), TAPVR, and IAA (1 case in each; 2.1%). The categories of arrhythmia were atrial flutter or fibrillation (33 cases; 70.2%), paroxysmal supraventricular tachycardia (6 cases; 12.8%), ventricular tachycardia (2 cases; 4.3%), and bradycardia (6 cases; 12.8%). The most common cardiac anomaly with atrial flutter-fibrillation was the post-Fontan surgery condition (23 cases; 69.7%). The most common reason for bradycardia was pacemaker malfunction (4 cases). None of the patients in this subgroup died during the hospitalization.

In the subgroup with signs of heart failure, the patient mean age was 7.6 ± 9.4 years and the median age was 6.75 years (range 1 month to 24.8 years). Among 18 cases (42.9%) in this subgroup, cardiac surgery was performed within four weeks of hospitalization. Their underlying cardiac lesions were VSD (13 cases; 31.0%), a complex univentricular heart (9 cases; 21.4%), DORV, TOF (7 cases in each; 16.7%), PDA, ECD (2 cases in each, 4.8%), and other (e.g., CoA and truncus arteriosus). Patients that had a TOF with heart failure were complicated by major aortopulmonary collateral arteries (MAPCAs) or the absent pulmonary valve syndrome. Two patients (4.8%) in this subgroup died within four weeks of hospitalization; one died due to sudden cardiac arrest with right ventricular failure, and the other due to ventricular tachycardia followed by sudden cardiac arrest. In 30 cases (71.4%) patients had already undergone previous cardiac surgery before the PER visits and in 14 cases (28.6%) the patients had no history of a previous cardiac operation. The causes of aggravated heart failure in the patients with previous surgical repair were pulmonary hypertension (15 cases; 50.0%), myocardial dysfunction (4 cases; 13.3%), aggravated valve regurgitation (3 cases; 10.0%), and other uncorrected lesions in 26.7%.

In the subgroup of patients with aggravated cyanosis, the mean age was 0.8 ± 1.4 years with a range from 4 days to 5.6 years. Their mean duration of hospitalization was 29.1 ± 20.0 days and in 11 patients (73.3%) in this subgroup cardiac surgery was performed within four weeks of hospitalization. Their symptoms of cyanosis were aggravated by shunt narrowing (or occlusion), hypoxic spells, aggravated pulmonary hypertension and
rupture of the baffling patch in 7, 4, 3, and 1 case, respectively. There were no deaths in this subgroup. Their underlying cardiac lesions were TOF (5 cases; 33.3%), TGA (4 cases; 26.7%), single ventricle or post-Fontan procedure (3 cases; 20.0%), CoA, critical PS, and ASD with pulmonary hypertension (1 case each).

**Discussion**

There are many studies reported in the medical literature on CHD lesions with regard to medical and surgical management. However, there is limited information on the ER presentation of patients with CHD. The goal of this study was to describe the presenting signs and symptoms of patients with CHD that presented to the ER and their clinical course.

We divided our study population into two major groups to identify the more common clinical patterns on presentation: the cardiopulmonary function might be compromised after long term sustained abnormal hemodynamics such as a shunt lesion and the inevitable changes of the hemodynamics after cardiac surgery. In group 2 patients with previously known CHD, surgery was performed less frequently within four weeks of hospitalization (74.3% in Group 1, 20.2% in Group 2). This may reflect the fact that other systemic morbidities increasingly effect and aggravate the cardiovascular condition of such patients. The non-surgical aggravation of cardiovascular function such as dysrhythmia, pulmonary hypertension, or myocardial dysfunction play an more important role in the long term follow up of these patients.

PGE1 infusion is essential for some newborn patients that depend on the patency of the ductus arteriosus for survival. Patients with ductus-dependent lesions presented to the ER with cardiac emergencies such as cyanosis and shock during the first few weeks of life. In our study, there were no deaths among patients with ductus-dependent circulation due to the immediate and timely PGE1 infusions. PGE1 infusion should be considered immediately in a critically ill infant presenting with acute cyanosis or severe systemic hypoperfusion, if ductus-dependent circulation is suspected.

Respiratory tract infection is a common manifestation of CHD, especially in early childhood; it is usually not considered to be a fatal disease among healthy children. In our study the mortality rate associated with respiratory tract infections, in patients with previous cardiac surgery, was 5.8%. There were many patients presenting with symptoms of respiratory tract infection that were discharged from the ER without hospitalization. Therefore only the more severely affected patients were included in our study population. However, these findings reflect the critical interaction between the cardiac and pulmonary systems. The younger patients were more susceptible to respiratory tract infections such as bronchiolitis and viral pneumonia. Furthermore, a subset of these patients had other concomitant conditions, such as malnutrition, and chromosomal or genetic abnormalities, which are associated with an increased susceptibility to infection, especially of the respiratory tract. The relatively high mortality rate indicates that common respiratory tract infections may compromise cardiac function and be fatal in this population.

In our study population, there were 48 cases over 20 years of age. Among these adulthood patients with CHD, 22 patients (45.8%) presented with symptoms of dysrhythmia. Recently, the adult population with CHD has been expanding due to advances in diagnostic modalities, open heart surgery, anesthesiology, and intensive care. With the increase in the number of survivors, the long term outcome of CHD among adults requires further investigations to understand the natural history of adult CHD and provide appropriate follow up and treatment. In particular, dysrhythmias have been identified as a major complication that can present in adults long after open heart surgery. Dysrhythmias frequently developed after the Fontan-type operation and atrial-switch, as well as after repair of TOF. The incidence of ventricular dysrhythmias in CHD patients is lower than for those with acquired cardiac disease. The dysrhythmias are thought to be due to atrial scar tissue in patients that have undergone repair at the atrial level. Atrial flutter was a common type of dysrhythmia in this study. Some studies have reported a positive correlation between the occurrence of atrial flutter and atrioventricular regurgitation in patients with CHD. The results of this study showed a strong relationship between the age of patients with CHD and the common clinical manifestations that are seen in the PER. In Group 2, the subgroups of respiratory tract infection and aggravated cyanosis were relatively younger than the subgroup of heart failure; the dysrhythmia subgroup was the oldest among the major clinical manifestations. This may be explained by the differences in susceptibility to infections, the natural course of CHD, and the influence of cardiac surgery.

The limitations of this study include the following. First, the retrospective study design, and exclusion of patients with CHD that were not admitted to the hospital. Therefore, the problems that did not require admission were not considered. Second, because most critically and acutely compromised neonates with CHD are frequently transferred directly to intensive care units without presenting to the PER, their information was not included in this study. For example, severely decompensated neonates with CHD such as TAPVR were commonly transferred to the neonatal intensive care unit of our hospital directly from other hospitals. This direct transfer to the intensive care unit may be a more
desirable mode of transfer for the safe and effective medical treatment in critically ill patients. In addition, some of the adult patients with CHD might have been missed because they presented to the adulthood ER, not the PER, and were admitted to the department of internal medicine.

Patients with CHD present with a variety of clinical symptoms to the ER. The symptoms of CHD might include multiple organ systems. Because of the complexity of the management of these patients, clinicians must be aware of the age-specific common manifestations, for prompt and accurate management of patients with CHD in the ER.

REFERENCES
1) Abu-Harb M, Hey E, Wren C. Death in infancy from unrecognized congenital heart disease. Arch Dis Child 1994;71:3-7.
2) Savitsky E, Alejos J, Votey S. Emergency department presentations of pediatric congenital heart disease. J Emerg Med 2003;24:239-45.
3) Watanabe M, Aoki M, Fujiwara T. Transition of ventricular function and energy efficiency after a primary or staged Fontan procedure. Gen Thorac Cardiovasc Surg 2008;56:498-504.
4) Silberbach GM, Imus RL, McDonald RW, Andrilenas K, Rice MJ, Reller MD. Effect of patent ductus arteriosus on Doppler-derived right ventricular systolic time intervals. Pediatr Cardiol 1993;14:155-8.
5) Howlett G. Lung mechanics in normal infants and infants with congenital heart disease. Arch Dis Child 1972;47:707-15.
6) Kaemmerer H, Bauer U, Pensl U, et al. Management of emergencies in adults with congenital cardiac disease. Am J Cardiol 2008;101:521-5.
7) Yee L. Cardiac emergencies in the first year of life. Emerg Med Clin North Am 2007;25:981-1008.
8) Woods WA, McCulloch MA. Cardiovascular emergencies in the pediatric patient. Emerg Med Clin North Am 2005;23:1233-49.
9) Medrano C, Garcia-Guereta L, Grueso J, et al. Respiratory infection in congenital cardiac disease: hospitalizations in young children in Spain during 2004 and 2005: the CIVIC epidemiologic study. Cardiol Young 2007;17:360-71.
10) Hilton JM, Fitzgerald DA, Cooper DM. Respiratory morbidity of hospitalized children with Trisomy 21. J Paediatr Child Health 1999;35:383-6.
11) Noh CI, Bae EJ. Atrial flutter in children. Korean Circ J 1991;21:107-16.