Heart Murmur in the First Week of Life: Siriraj Hospital

Duangmanee Laohaprasitiporn MD*, Tipawan Jiarakamolchuen MD*, Prakul Chanthong MD*, Kritvikrom Durongpisitkul MD*, Jarupim Soongswang MD*, Apichart Nana MD*

*Department of Pediatrics, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand

Congenital heart disease (CHD) is the most common form of cardiovascular diseases in children. In Thailand, there has not been available information about congenital heart disease in neonates. Between January 1st and December 31st, 2000, all full-term babies born at Siriraj Hospital with detected heart murmur within the first week of life were consulted to pediatric cardiologists. Echocardiography was performed for diagnosis in every baby. Total livebirths during that period were 11,245 cases. Heart murmurs were detected in 83 cases. The incidence of heart murmur within the first week of life was 7.38:1,000 livebirths. Innocent murmurs were found in 34 cases and echocardiogram revealed no detectable cardiac anomalies (2 cases), mild tricuspid regurgitation (2 cases), physiologic branch pulmonary stenosis (4 cases), and small size PDA (≤ 2 mm., 26 cases). Forty-nine cases had CHDs. The incidence of CHD was 4.36:1,000 livebirths. At the time of initial diagnosis, 22 cases (44.8%) were asymptomatic. Among these patients, 1 case had serious cardiac anomaly, i.e., tetralogy of Fallot. There were 27 cases with symptoms, including 15 cases (30.6%) with tachypnea, 8 cases (16.4%) with cyanosis and 4 cases (8.2%) with congestive heart failure. The 3 most common cardiac diseases were ventricular septal defect (9 cases, 18.4%), patent ductus arteriosus greater than 2 mm. (8 cases, 16.3%), and atrial septal defect (8 cases, 16.3%). Those with CHDs were treated with anticongestive medications (22 cases, 44.8%), prostaglandin E, (5 cases, 10.2%), laser pulmonary vulvulotomy (1 case, 2%), palliative surgery within the first week of life (4 cases, 8.2%) and corrective surgery (4 cases, 8.2%). During follow-up for the period of 1 year, 2 cases died from sepsis. Early diagnosis and proper management are important to reduce morbidity and mortality in the newborn with CHD.

Keywords: Heart murmur, congenital heart disease, newborn

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The incidence of congenital heart disease (CHD) worldwide is rather constant, occurring at 8-10:1,000 livebirths⁽¹⁾. Some malformations are not easily detected during neonatal period, some become modified or disappear during infancy and childhood. Several can cause death in the first few weeks or months of life if they are not recognized⁽²⁾. Twenty five per cent of the Swedish and 29 per cent of the American babies with CHD died in the first year of life, mostly (61%) in the neonatal period⁽²⁾. In 2001, we reported CHD in infants at Siriraj Hospital: 51 cases (30.9%) born in our hospital were diagnosed as

having CHD before discharge. During neonatal period, acyanotic congenital heart disease was found more common (76.2%) than cyanotic patients (27.3%). Asymptomatic patients presented with heart murmur was found in $38.1\%^{(3)}$.

Innocent murmur can be detected in the first week of life in approximate 1.7-7.7%⁽⁴⁾. Since there has been no detail about CHD and innocent murmur in this age group, the purpose of the present study was to investigate the etiology of heart murmur detected within the first week of life in all full-term babies born at Siriraj Hospital.

Material and Method

All full-term babies (>37 weeks gestational age) born at Siriraj Hospital between January and

Correspondence to: Duangmanee Laohaprasitiporn, MD, Department of Pediatrics, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok 10700, Thailand Telephone: 0-2419-7000 ext. 5932, 5672 Facsimile: 0-2411-3010 e-mail: sidlh@mahidol.ac.th

December 2000 with detectable heart murmur within the first week of life were enrolled in the present study. The diagnosis was confirmed by two-dimensional imaging and color Doppler echocardiography in all cases. Some patients required cardiac catheterization for further management. Congenital heart diseases are defined as presence of a gross structural abnormality of the heart or intrathoracic great vessels that is functional significance⁽⁵⁾. Innocent murmur is diagnosed when heart murmur has been detected without symptoms and with normal echocardiographic findings or with no significant structural heart defect^(4,6), i.e., tiny patent ductus arteriosus (diameter less than 2 mm.) which can be considered abnormal if remains patent after 3 months⁽⁷⁾, physiologic branch pulmonary stenosis of the newborn⁽⁸⁾ or mild tricuspid regurgitation. Demographic data, i.e., sex, birth weight, age at audible heart murmur, associated extracardiac anomaly, clinical manifestations, types of heart murmur, types of cardiac malformation by echocardiographic findings and management were recorded.

Statistical analysis

The categorical variables were presented as count and percentage. The continuous variables are reported as mean \pm SD.

Results

Between January and December 2000, there were total of 11,245 full-term livebirths at Siriraj Hospital. Eighty three babies, aged within the first week of life who presented with heart murmur were evaluated by pediatric cardiologists. It accounted for the incidence of audible heart murmur of 7.38:1,000 livebirths. Thirty nine cases (46.9%) were males and 44 cases (53.1%) were females. The mean birth weight was $3,016.7\pm67$ gm (range, 2,410-4,530 gm). Heart

murmur was detected at the mean age of 27.7 ± 0.4 hours. The murmur was found within the first 3 days of life in 76 cases (91.6%) and between 4-7 days of life in 7 cases (8.4%). All heart murmurs were graded less than 3/6 (Table 1).

Diagnosis

All babies had two-dimensional echocardiography and color Doppler mapping performed. The findings were categorized into 2 groups.

1. Congenital structural heart defects were found in 49 cases (59%), accounting for the incidence of CHD 4.36:1,000 livebirths. There were 23 males (46.9%) and 26 females (53.1%) in this group. Single anomaly was demonstrated in 25 cases (51%) and more than 2 anomalies in 24 cases (49%) (Table 2, 3). The diagnosis of multiple cardiac lesions was classified according to the dominant lesion, based on its physiologic importance.

2. Normal or no significant cardiac anomaly, described as innocent murmur were found in 34 cases (41%). These included normal structural heart 2 cases (5.9%), physiologic branch pulmonary stenosis (PS) 4 cases (11.7%), mild tricuspid regurgitation 2 cases (5.9%) and tiny PDA 26 cases (76.5%).

Associated extracardiac anomalies in the patients with CHD

Down syndrome was found in 6 cases (12.3%). The associated CHDs were ventricular septal defect/PDA/atrial septal defect (VSD/PDA/ASD) 1 case, PDA 3 cases, PDA/ASD 1 case, and common atrium/common ventricle 1 case. Cleft lip associated with PDA/ASD/VSD was found in 1 case. Cleft lip with holopro-encephaly was found in a patient with common atrium/common ventricle.

Table 1	. Types,	grading,	locations and	causes of heart	murmur in the	e present study	(n = 83)
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Heart murmur			Cases	Diagnosis by	
Туре	Grading	Location	(%)	echocardiogram	
Pansystolic	2	Left lower sternal border	6 (7.2)	VSD (4 cases), TR (2 cases)	
Ejection systolic	1-2	Left lower sternal	7 (8.3)	VSD (5 cases), normal (2 cases)	
Continuous	2	Left upper sternal border	2 (2.5)	PDA (2 cases) Innocent (30 cases).	
Ejection systolic	2-3	Left upper sternal border	68 (82)	CHD (38 cases)	

VSD, ventricular septal defect; TR, tricuspid regurgitation; PDA, patent ductus arteriosus; CHD, congenital heart disease

Clinical manifestation

There were 22 (44.8%) asymptomatic neonates. Tachypnea was found in 15 cases (30.6%), cyanosis in 8 cases (16.4%) and congestive heart failure in 4 cases (8.2%).

Initial investigations in CHD group

Chest roentgenography was normal in 39 cases (79.6%) and abnormal in 10 cases (20.4%). The abnormal findings were (a) cardiomegaly and increased pulmonary blood flow 4 cases (40%; VSD/PDA/ASD 3 cases, VSD/PDA 1 case); (b) decreased pulmonary blood flow 5 cases (50%; pulmonary atresia (PA) with VSD/PDA, PA with intact ventricular septum (IVS), tricuspid atresia (TA) with PA/ASD, common ventricle/ subvulvular PS/PDA and common atrium/common ventricle/PA, 1 case in each cardiac lesion) and (c) dextrocardia with corrected TGA (1 case; 10%).

Electrocardiography was normal in 37 cases (75.5%). The abnormal findings in 12 cases (24.5%) were chamber enlargement 7 cases (58.4%) and abnormal axis 5 cases (41.6%).

Management

There were 27 patients with CHD (55.1%) who required medications, i.e., anticongestion 22 cases (44.8%), and prostaglandin E_1 5 cases (10.2%). Cardiac catheterization intervention with laser pulmonary valvulotomy was performed in 1 patient with PA/IVS. Surgical treatment was performed in 8 cases (16.3%), i.e., palliative surgery 4 cases and corrective surgery 4 cases. The operation included modified Blalock-Taussing shunt, PDA ligation, Dacron patch VSD closure and coarctectomy. The age at operation varies from 1 week to 6 months. The other 22 cases (44.9%) required neither medication nor intervention.

Complications

Three patients (6.1%) with CHD developed complications. All of them were non cardiac causes; renal failure with recovery 1 case, and death 2 cases (unrepaired TGA 1 case and unrepaired TOF 1 case) from sepsis and meningitis.

Discussion

The incidence of congenital heart disease (CHD) has been remarkedly constant worldwide and

Table 2. Types of congenital heart diseases (CHD) in the present study (n = 49)

Types of CHD	No., cases (%)		
VSD	9	(18.4)	
PDA	8	(16.3)	
ASD	8	(16.3)	
PDA/ASD	5	(10.3)	
Common atrium/common ventricle	4	(8.3)	
VSD/PDA/ASD	4	(8.3)	
VSD/PDA	3	(6.1)	
Common ventricle/subvulvular PS/PDA	1	(2.0)	
Corrected TGA	1	(2.0)	
DTGA/VSD/ASD/PDA	1	(2.0)	
PA/IVS	1	(2.0)	
PA/VSD/PDA	1	(2.0)	
TA/PA/ASD	1	(2.0)	
TOF	1	(2.0)	
VSD/Coarctation of aorta/PDA	1	(2.0)	

VSD, ventricular septal defect; PDA, patent ductus arteriosus; ASD, atrial septal defect; TGA, transposition of the great arteries; PA, pulmonary atresia; IVS, intact ventricular septum; PS, pulmonary stenosis; TA, tricuspid atresia; TOF, tetralogy of Fallot

Table 3. Types of congenital heart diseases (CHD) in the present study compared to the other studies which included the patients aged under 15 years old. (presented as percentages)

CHD	Siriraj 2000 (n = 49)	Multicenter ⁽⁹⁾ 1993 (n = 442)	New England ⁽¹⁰⁾ 1977 (n = 2,381)	Chula ⁽¹¹⁾ 1992 (n = 345)	Bhurirum ⁽¹²⁾ 1992-97 (n = 339)	Children ⁽¹³⁾ 1987-1992 (n = 1,953)	Rama ⁽¹⁴⁾ 1990-1994 (n = 1,408)
VSD	18.4	38.5	15.7	36.2	22.1	20.3	28.1
PDA	16.3	17.4	6.1	10.7	16.1	15.4	15.2
TOF	2.0	8.4	8.9	11.3	11.2	14.9	11.3
ASD	16.3	5.1	2.9	4.6	5.2	7.0	6.1
PS	10.2	6.5	3.3	5.8	4.5	4.1	8.7
Other	36.8	24.1	36.9	43.1	40.9	38.3	30.6

Chula = Chulalongkorn Hospital; Rama = Ramathibodi Hospital

over the years $^{(1,15)}$. In the present study, we found the incidence of congenital heart disease in full-term babies of 4.36:1,000 livebirths. The reasons of lower incidence in our report were possibly due to 1) the narrow range of age recruited in our study (within 1st week of age), which some malformations are not easily detected soon after birth, 2) physical examination in the majority of our cases that were performed by the first or second year resident trainees. The distribution of lesions and age at presentation in the present study were similar but not identical to the previous studies. However, the distribution of major lesions were significantly related to the age at presentation. In the first week of life, the most common cardiac lesion was transposition of the great arteries, followed by hypoplastic left ventricle, tetralogy of Fallot and coarctation of the aorta⁽⁸⁾. In the present study, the 3 most common CHDs were VSD, PDA and ASD. None of the cases in this study was diagnosed as having aortic stenosis, perhaps due to a much lower incidence in the Asian than the Western population⁽¹⁶⁾. Fifty-five per cent of our patients were symptomatic within the first week of age, similar as in the other reports^(17,18). Cardiogenic shock had been reported in less than $1\%^{(9)}$. We did not have any patients with this symptom, even though symptoms of congestive heart failure were found in 44.9%. Ten per cent of our patients required prostaglandin E₁; all of them had ductus-dependent pulmonary circulation lesions. Mortality rate of CHD in the present study was 4.1% which was same as the other report $(3-4\%)^{(17)}$. However, the very high mortality rate from severe congenital cardiac malformation was found in the first week of life (9-32%)^(19,20). This emphasizes the importance of early detection. If lives are to be saved, a diagnosis must be made and treatment should be initiated as soon as possible after birth. In the present study, 55% of the patients required life-saving medications. Approximately 20% required cardiac catheterization intervention and operation; the figure which was higher than in other report $(12.5\%)^{(9)}$. All neonates should be carefully examined before discharge from hospital, or soon after birth if born outside hospital by a pediatrician and referred to a cardiologist if a cardiac anomaly is suspected⁽²⁾.

Chest roentgenography and electrocardiography are used as initial tools in diagnosis CHD. In the present study, the majority of CHD patients had normal results with normal chest roentgenogram in 79.6%, and normal electrocardiogram in 75.5%. So every neonate who has been suspected to have CHDs, especially those who become symptomatic, should undergo two-dimensional imaging and color Doppler echocardiography as soon as possible. Innocent murmur in the first week of life had been reported $1.7-7.7\%^{(16)}$, but there was only 0.3% in the present study. The associated extracardiac anomalies was found in 16.3% of CHD patients, which was same as the other report (14.8-20%)^(11,14).

Conclusion

Since congenital heart disease is relatively rare in general population, screening with echocardiography is difficult to apply in every neonate, especially in Thailand due to high cost and limited supply of pediatric cardiologists. Clinical assessment should be carefully performed in every neonate. All suspected patients especially with specific signs and symptoms, such as having clues of serious cardiovascular anomaly, i.e., early congestive heart failure and/or cyanosis should be stabilized and referred for early definite diagnosis and proper management to reduce morbidity and mortality.

Limitations in this study were: 1) rather small number of subjects due to the short period of the study (only 1 year). Extended study including long-period followup should be considered. 2) limitation in skill of physicians to detect heart murmur.

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เสียงหัวใจผิดปกติในทารกอายุ 1 สัปดาห์แรก

ดวงมณี เลาหประสิทธิพร, ทิพวรรณ เจียรกมลชื่น, ประคัลภ์ จันทร์ทอง, กฤตย์วิกรม ดุรงค์พิศิษฏ์กุล, จารุพิมพ์ สูงสว่าง, อภิชาติ นานา

้โรคหัวใจพิการแต่กำเนิดเป็นสาเหตุของโรคหัวใจและหลอดเลือดที่พบมากที่สุดในเด็ก เนื่องจากยังไม่เคยมี รายงานเกี่ยวกับเสียงหัวใจที่ฟังได้ผิดปกติในทารกแรกเกิดในโรงพยาบาลศิริราช จึงได้ทำการศึกษาในโรงพยาบาล ้ศิริราช ตั้งแต่วันที่ 1 มกราคม ถึง 31 ธันวาคม 2543 โดยทารกคลอดครบกำหนดทุกรายที่มีอายุภายในสัปดาห์แรก ซึ่งมีเสียงหัวใจผิดปกติและส่งปรึกษาหน่วยโรคหัวใจเด็ก ภาควิชากุมารเวชศาสตร์ จะได้รับการตรวจหัวใจด้วย คลื่นเสียงสะท้อนความถี่สูง เพื่อยืนยันการวินิจฉัย จากการศึกษาพบว่า มีจำนวนทารกแรกเกิดที่คลอดมีชีพ ในช่วงเวลาดังกล่าว 11,245 ราย ฟังเสียงหัวใจผิดปกติ 83 ราย คิดเป็นอุบัติการณ์ 7.38:1,000 รายของทารกคลอด ี่มีชีพ มีทารกจำนวน 34 ราย ที่วินิจฉัยว่าเป็น innocent murmur โดยไม่พบความผิดปกติของโครงสร้างหัวใจ 2 ราย ลิ้นหัวใจไตรคัสปิดรั่วเล็กน้อย 2 ราย เป็น physiologic branch pulmonary stenosis 4 ราย และมี patent ductus arteriosus ขนาดเล็กกว่า 2 มิลลิเมตร 26 ราย พบทารกที่เป็นโรคหัวใจพิการแต่กำเนิด 49 ราย คิดเป็นอุบัติการณ์ โรคหัวใจพิการแต่กำเนิดที่ตรวจพบในทารกอายุสัปดาห์แรกในโรงพยาบาลศิริราช 4.36:1,000 รายของทารกคลอด มีชีพ โดยพบว่าทารกที่ไม่มีอาการแสดงและฟังได้เสียงหัวใจผิดปกติมี 22 ราย (ร้อยละ 44.8) ซึ่งในจำนวนนี้มี 1 ราย ้(ร้อยละ 4.5) ได้รับการวินิจฉัยว่าเป็น tetralogy of Fallot ทารกที่มีอาการผิดปกติได้แก่ อาการหายใจเร็ว 15 ราย (ร้อยละ 30.6) อาการเขียว 8 ราย (ร้อยละ 16.4) และอาการหัวใจวาย 4 ราย (ร้อยละ 8.2) โรคหัวใจพิการแต่กำเนิด ที่พบมากที่สุด 3 ชนิดเรียงตามลำดับจากมากไปน้อยคือ ventricular septal defect 9 ราย (ร้อยละ 18.4), patent ductus arteriosus ที่มีขนาดใหญ่กว่า 2 มิลลิเมตร 8 ราย (ร้อยละ 16.3) และ atrial septal defect 8 ราย (ร้อยละ 16.3) ทารกที่เป็นโรคหัวใจพิการแต่กำเนิดในการศึกษานี้ ได้รับการรักษาด้วยยาควบคุมภาวะหัวใจวาย 22 ราย (ร้อยละ 44.8) และ prostaglandin E_. 5 ราย (ร้อยละ 10.2) รักษาทางสายสวนหัวใจ 1 ราย (ร้อยละ 2) รักษาด้วยการ ผ่าตัด 8 ราย (ร้อยละ 16.3) เป็น palliative surgery 4 ราย (ร้อยละ 8.2) ซึ่งทุกรายได้รับการผ่าตัดในช่วงอายุสัปดาห์ แรก และทำ corrective surgery 4 ราย (ร้อยละ 8.2) จากการติดตามทารกที่เป็นโรคหัวใจพิการแต่กำเนิดภายใน ระยะเวลา 1 ปี พบว่ามี 2 รายที่เสียชีวิตจากสาเหตุอื่นคือภาวะติดเชื้อ ดังนั้นการให้การวินิจฉัยโรคหัวใจพิการแต่กำเนิด ้อย่างทันท่วงที่และให้การรักษาอย่างเหมาะสมและรวดเร็วจะช่วยลดภาวะแทรกซ้อนและอัตราการตายในทารกเหล่านี้