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Epidemiological Profile of Congenital Heart Disease in A National Referral Hospital

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Abstract

Introduction: Sardjito Hospital is the tertiary referral hospital in Yogyakarta and Central Java, Indonesia. There are no reported data measuring epidemiological profile of congenital heart disease (CHD) in this region. This study was aimed specifically to determine the most major lesion and type of CHD in one of an over-populated area in Indonesia.

Methods: The retrospective observational study was conducted during January 2014 to December 2014 in Sardjito Hospital. In and outpatients with CHD taken from medical record were noted their clinical data and only patients with transthoracic echocardiographic proof of CHD were included in this study. CHD with more than single lesion was simplified as one lesion which had most major impact on hemodynamic circulation. The denominator was the sum of all new patients visiting to the hospital at 2014. Data were entered in MS-excel and analyzed by software SPSS version 22.

Results: We had total 650 new patients with CHD registered to Sardjito Hospital in 2014. The incidence of CHD was 134/10.000 person-years consisted of adult 22% and children 78% (female 60% and male 40%). Ventricle septal defect (VSD) was the most common lesion among children (30%) followed by atrial septal defect (ASD) 17%, persistent ductus arteriosus (PDA) 16%, and Tetralogy of Fallot (TOF) 7%. Meanwhile in adult, ASD was the most frequent CHD (60%), continued by VSD 23%, TOF 8%, and PDA 4%. Secundum, primum, and sinus venosus type were found in 94%, 3%, and 3% of total ASD in children, and 99%, 1%, and 0% of adult respectively. Perimembran outlet (PMO), doubly committed subarterial (DCSA), and inlet were found in 66%, 12%, and 12% of total VSD in children, and 39%, 54%, and 0% of adult respectively. TOF had the highest incidence of cyanotic heart diseases (33 cases, 7%) in children and also in adult (12 cases, 8%).

Conclusion: The incidence of CHD was 134/10.000 person-years. The most common lesion among children was VSD and in adult was an ASD. TOF had the highest incidence of cyanotic heart disease among children and adult. Secundum type was the most common finding in all ages of ASD population, whereas PMO and DCSA were the most frequent type of VSD in children and adult respectively.

Keywords: incidence, congenital heart disease, hospital, cyanotic, type.

Introduction

Congenital heart disease (CHD) are serious and common conditions that have significant impact on morbidity, mortality, and healthcare costs in children and adults. Congenital heart disease refers to structural abnormality of the heart and or great vessels that is present at birth and has potential functional significance. It usually excludes congenital arrhythmias and cardiomyopathies even though these may be based on genetic or other abnormalities that are present at birth.¹ Moss and Adams' define CHD as the developmental changes of the intracardiac structures and major vessels and is interchangeable with the term cardiovascular malformation.²

Congenital heart disease is the most common cause of major congenital anomalies, representing a major global health problem. Reported birth incidence of CHD varies widely among studies worldwide. The reported incidence of CHD in the United States is between 4 and 10 per 1,000, clustering around 8 per 1,000 live births. Continental variations in birth prevalence have been reported, from 6.9 per 1000 births in Europe to 9.3 per 1000 in Asia.³

Patients with CHD often need long-term expert medical care and high cost of healthcare resources. The advances in diagnosis and surgical treatment over the past 40 years have led to dramatic increases in survival for children with serious heart

defects in Indonesia.⁴ Most patients born with CHD now have reached adulthood. As a result, the population of adult CHD has been steadily growing. However, the size of the current population of adults with CHD is unknown.

The global health burden as the result of CHD increases quickly. To assess the needs for care for these patients, we need first to determine the incidence of CHD. In 2004, the total hospital costs for congenital cardiovascular defect conditions was around \$2.6 billion. The highest costs were for stays related to cardiac and circulatory congenital anomalies, which accounted for ≈\$1.4 billion, more than half of all hospital costs for birth defects. In 2009, 52,000 U.S. adults and children (25,000 males; 27,000 females) diagnosed with CHD, were discharged from short-stay hospitals.³

Webb and Bain mention that the incidence of disease measures how quickly people are catching the disease and it differs from prevalence because it considers only new cases, that occurred in a specific time period.⁵ The prevalence of a disease tells us what proportion of a population actually has the disease at a specific point in time.⁶

Incidence refers to the number of new affected persons per unit of time or population; prevalence refers to the number present at any time and represents the difference between the incidence and those who die in the same period. The incidence of CHD is usually estimated by calculating the number of subjects with CHD per thousand or per million live births.¹

Sardjito Hospital is a tertiary referral hospital which was selected by Indonesian Ministry of Health as one of the national referral hospitals in DI Yogyakarta and south area of Central Java provinces since 2014. The population of DI

Yogyakarta province in 2010 was 3.457.491 lives. Meanwhile in south area of Central Java province (Boyolali, Klaten, Magelang, and Purworejo regency) was resided by 3.937.728 people.⁶ There are no reported data measuring epidemiological profile of CHD in this region. This study aimed specifically to determine the most major lesion and type of CHD in Indonesia, using hospital-based sample.

Methods

The retrospective observational study was conducted during January 2014 to December 2014 in Sardjito Hospital. In and outpatients CHD clinical data were taken from medical records data and only new patients who were firstly diagnosed by transthoracic echocardiography proof of CHD were included in this study. Patients with grown up CHD after surgical or non surgical correction were excluded. CHD with more than single lesion was simplified as one lesion which had most major impact on hemodynamic circulation. The denominator was the sum of all new patients visiting to the hospital at 2014. Data were entered in MS-excel and analyzed by software SPSS version 22.

Results

The age of children patient ranged from 0 days to 17 years ($2,7 \pm 4,4$ yo) and adult patient ranged from 18 to 70 years ($32,7 \pm 13,4$ yo). Among the 48.480 new patients (out and inpatients) registered to Sardjito Hospital in 2014, there were 650 patients with CHD. The incidence of CHD was 134/10.000 person-years consisted of adult 3/1000 person years, children 10.4/1000

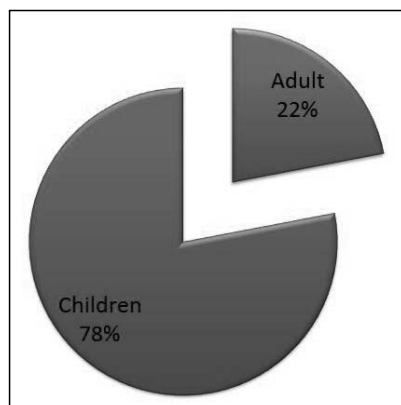


Figure 1. Incidence of CHD in Sardjito Hospital at 2014 (age based-in percentage)

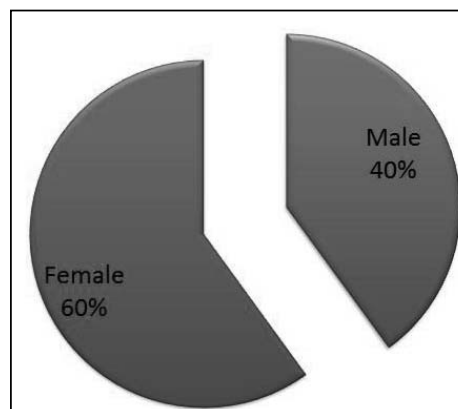


Figure 2. Incidence of CHD in Sardjito Hospital at 2014 (gender based-in percentage)

person years and female 8/1000 person years, male 5.4/1000 person years. Children patients accounted for 78% of the CHD population and 22% in adults (Figure 1). Gender based percentage showed 60% for female and 40% male (Figure 2).

Ventricle septal defect (VSD) was the most common lesion among children (152 patients,30%) followed by atrial septal defect (ASD) 86 patients (17%), persistent ductus arteriosus (PDA) 81 patients (16%), and Tetralogy of Fallot (TOF) 33 patients (7%)(Table 1). Meanwhile in adult, ASD was the most frequent CHD (87 patients,60%), followed by VSD 33 patients (23%), TOF 12 patients(8%), and PDA 6 patients(4%) (Table 2).The highest incidence of cyanotic heart diseases was TOF (33 cases, 7%) in children and also in adult (12 cases, 8%).

Secundum, primum, and sinus venosus type were found in 94%, 3%, and 3% of total ASD

in children (Figure 3), and 99%, 1%, and 0% of adult respectively (Figure 4). Perimembran outlet (PMO), doubly committed subarterial (DCSA), and inlet were found in 66%, 12%, and 12% of total VSD in children (Figure 5), and 39%, 54%, and 0% of adult respectively (Figure 6).

Discussion

An estimated 150 million live births per year worldwide and 1.35 million were affected with CHD. Around 85% of the world’s population lives in low or middle-income countries (developing countries), where considerable challenges limited access to the appropriate CHD healthcare.⁷ There was lack of data in the incidence or birth prevalence of CHD in most developing countries, so that the impacts of these defects have been systematically underestimated in the past. For these reasons, prevalence studies though more readily available

Table 1. Type and Proportion of Congenital Heart Disease in Children

NO	TYPE OF CHD	NUMBER OF CASES (%)	NO	TYPE OF CHD	NUMBER OF CASES (%)
1	VSD	152 (30,2)	15	SINGLE VENTRICLE	3 (0,6)
2	ASD	86 (17,1)	16	SINGLE ATRIUM	2 (0,4)
3	PDA	81 (16,1)	17	TRUNCUS ARTERIOSUS	2 (0,4)
4	TOF	33 (6,6)	18	ATRESIA MITRAL	1 (0,2)
5	PS	29 (5,8)	19	ATRESIA TRIKUSPID	1 (0,2)
6	PFO	26 (5,2)	20	COARTASIO AORTA	1 (0,2)
7	ATRESIA PULMONAL	19 (3,8)	21	CC TGA	1 (0,2)
8	DORV	16 (3,2)	22	DOLV	1 (0,2)
9	AVSD	15 (3,0)	23	ECTOPIA CORDIS	1 (0,2)
10	TGA	13 (2,6)	24	MS	1 (0,2)
11	DEKSTROCARDIA	6 (1,2)	25	PAPVD	1 (0,2)
12	PENTALOGY OF FALLOT	5 (1,0)	26	TAPVD	1 (0,2)
13	EBSTEIN ANOMALY	3 (0,6)	27	HCM	1 (0,2)
14	AORTA STENOSIS	3 (0,6)		TOTAL	504 (100)

Note:VSD = ventricular septal defect; ASD = atrial septal defect; PDA = patent ductus arteriosus; TOF = tetralogi of fallot; PS = pulmonary stenosis; PFO = patent foramen ovale; DORV = double outlet right ventricle; AVSD = atrioventricular septal defect; TGA = transposition of the great arteries; CC = congenitally corrected; DOLV = double outlet left ventricle; MS = mitral stenosis; PAPVD = partial anomalous pulmonary venous drainage; TAPVD = total anomalous pulmonary venous drainage; HCM = hyperthropic cardiomyopathy.

Table 2. The Type and Proportion of Congenital Heart Disease in Adult

NO	TYPE OF CHD	NUMBER OF CASES (%)
1	ASD	87 (60)
2	VSD	33 (23)
3	TOF	12 (8)
4	PDA	6 (4)
5	PFO	2 (1)
6	SINGLE ATRIUM	2 (1)
7	PS	1 (1)
8	AP WINDOW	1 (1)
9	CCTGA	1 (1)
10	DEKSTROCARDIA	1 (1)
	TOTAL	146 (100)

Note: ASD = atrial septal defect; VSD = ventricular septal defect; TOF = tetralogi of fallot; PDA = patent ductus arteriosus; PFO = patent foramen ovale; PS = pulmonary stenosis; AP = aorto-pulmonary; CCTGA = congenitally corrected transposition of the great arteries.

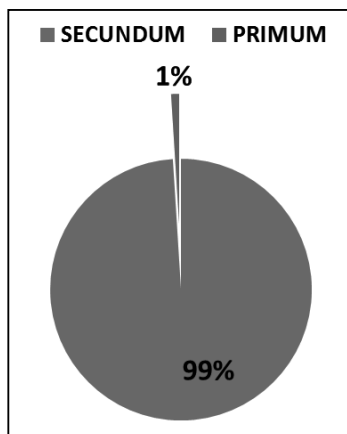


Figure 3. Type of ASD in Adult

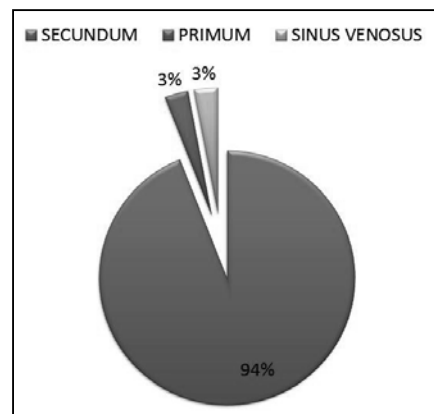


Figure 4. Type of ASD in Children

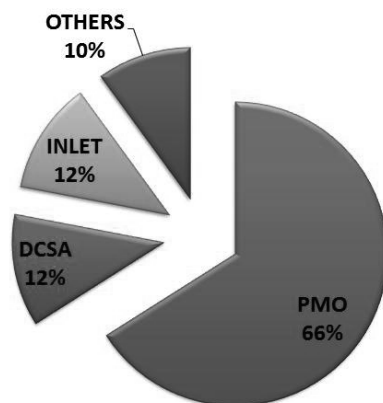


Figure 5. Type of VSD in Children. PMO = perimembrane outlet; DCSA = doubly committed subarterial

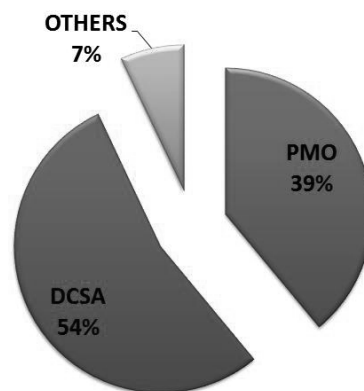


Figure 6. Type of VSD in Adult. PMO = perimembrane outlet; DCSA = doubly committed subarterial

in developing countries than incidence studies, tend to underestimate the burden of congenital heart disease.⁸

The incidence of CHD in our study was a hospital-based not a community based study. A comparison with another hospital-based study in Nepal by Shah *et al* showed that the incidence of CHD in our study was 1.8 times higher, despite the discrepancy in the study subjects.⁹ Shah *et al* (2008) reported incidence of CHD in 2006 among 14,461 hospitalized children was 5.8 per 1000.⁹ A hospital based study in India which was conducted in 1994 showed the prevalence of CHD among 10,964 live birth was 39 per 10,000.⁸

Ventricular septal defect (VSD) was the most common form of congenital heart defect in children, accounted for 30% of total CHD, followed by atrial septal defect (ASD, 17%), patent ductus arteriosus (PDA, 16%), and tetralogy of fallot (TOF, 7%) . Perimembranous type (PMO) of VSD was the most frequent type (66%). This result similar with the previous study who stated that VSD was accounted for 20% of total CHD and 80% of these were perimembranous type.¹⁰ Atrial septal defect was the most frequent defect CHD in adult (60%), followed by VSD (23%), and TOF (8%). It was similar to the study from Hannoush *et al* who stated that ASD was the most common CHD (53%), followed by VSD (11%) dan TOF (11%). The higher relative frequency of ASD may be related to failure in early detection, since these patients are usually asymptomatic during childhood.¹¹

The disease burden can also be estimated by comparing the proportion of adults to children with CHD. The burden of CHD by WHO region indexed to regional population by age using a CHD prevalence of 12/1000 before and 4/1000 after age 18 years.⁷ Marelli *et al* (2014) published the Quebec's CHD database which contained data on 107,559 individuals with CHD from 1983-2010. Overall prevalence of CHD in 2010 was 13.1/1000 in children and 6.12/1000 in adults.¹² By 2010, adults accounted for 66% of the entire CHD population.¹² A study in Newcastle between 1985 and 1994 noted 1,942 cases of CHD in a population of 377,310 livebirths (incidence of 5.2/1,000). Of these newborns, 1,514 were predicted to survive >16 years.¹³ In our hospital-based study, it was noted that the incidence of CHD was 10.4/1000 in children and 3/1000 in adults, and 22% of all patients were adults (GUCH were excluded).

Conclusion

The incidence of CHD was 134/10,000 person-years. The most common lesion among children was VSD and in adult was an ASD. TOF had the highest incidence of cyanotic heart disease among children and adult. Secundum type was the most common finding in all ages of ASD population, whereas PMO and DCSA were the most frequent type of VSD in children and adult respectively. Our findings informed that there was a high burden and the need to plan and provide healthcare resources.

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