

Pediatric heart diseases in Madina, Saudi Arabia

Current status and future expectations

Abdulhameed A. Alnajjar, MBBCh, MD, Mohamed-Mofeed F. Morsy, MSc, MD, Ibrahim S. Almuzainy, MBBCh, MD, Abdulmateen A. Sheikh, MBBCh, MD.

ABSTRACT

الأهداف: إبراز حجم مشكلة أمراض قلب الأطفال بالمدينة المنورة و كذلك الوضع الحالي للخدمة المقدمة مع إظهار العقبات و التوقعات المستقبلية لتلك الخدمة.

الطريقة: أجريت دراسة عرضية خلال الفترة ما بين يناير 2007 و حتى يونيو 2008 - مستشفى الولادة و الأطفال - المدينة المنورة - المملكة العربية السعودية. و قد تم اعتماد الدراسة من خلال اللجنة المنظمة للبحوث و أخلاق المهنة بالمستشفى. و لقد تم عمل برنامج كمبيوتر خاص لتسجيل بيانات جميع المرضى الذين ترددوا على وحدة قلب الأطفال خلال تلك الفترة.

النتائج: لقد تم فحص 4348 طفل خلال تلك الفترة و ذلك تضمن 2301 (53%) مريض من حالات المتابعة. و من بين 2047 طفل جديد تم فحصهم وجد أن هناك 794 (38.8%) طفل يعاني من أمراض في القلب. و لقد شكلت عيوب القلب الخلقية CHD 705 حالة من تلك الحالات، بينما شكل مرض اعتلال عضلة القلب 34 حالة، و أمراض القلب الروماتيزمية 26 حالة و ارتخاء الصمام المترالي 15 حالة، و مرض كاواساكي 9 حالات و اعتلال ضربات القلب 5 حالات. و لقد كان الثقب بين البطينين (34.5%) أكثر العيوب الخلقية شيوعا متبوعا بالثقب بين الأذنين (8.9%). و لقد كانت نسبة عيوب القلب الأخرى كالتالي: ضيق الشريان الرئوي (7.9%)، بقاء الوصلة الشريانية (6%)، الثقب ما بين الأذنين و البطينين (3.8%)، ضيق الشريان الأورطي (3.5%)، تناظر شرايين القلب (3.5%)، رباعي فالوت (3%)، ضيق برزخ الأورطي (2.8%) و العيوب الأخرى (26%). و من بين كل المرضى فلقد تم نقل 310 مريض إلى مراكز القلب بالرياض لإجراء قسطرة قلبية أو جراحة قلب. و كان معظم هؤلاء المرضى من ذوى العيوب الخلقية. و لقد تم نقل جميع المرضى عن طريق الجو بواسطة الطيران. و من بين هؤلاء المرضى تم نقل 128 مريض بصفة عاجلة متضمنا 66 مريضا تم نقلهم بواسطة طائرة الإخلاء الطبي.

خاتمة: تمثل أمراض القلب عند الأطفال مشكلة صحية كبيرة. و معظم هؤلاء المرضى يعانون من عيوب خلقية بالقلب CHD. و لقد تم التنويه على أهمية وجود مركز قلب بالمدينة المنورة لتفادي المشكلات الناجمة عن نقل مرضى القلب.

Objectives: To demonstrate the magnitude of pediatric heart disease problem in Madina, current situation of heart service, obstacles and future expectations.

Methods: We conducted this cross sectional study in Madina Maternity and Children Hospital, Madina, Kingdom of Saudi Arabia from January 2007 to June 2008. The study was approved by the Ethical Committee. A computer based program was established to register the data of all children attended the pediatric cardiology unit.

Results: A total of 4348 children were seen in the study period; 2301 were follow up cases while 2047 were new cases. Of the new cases, 794 (38.8%) were found to have heart diseases. Of this, 705 have CHD (cardiomyopathies 34, rheumatic heart disease 26, mitral valve prolapse 15, Kawasaki disease 9, and arrhythmia 5). Ventricular septal defect was the most common CHD (34.5%), and followed by atrial septal defect (8.9%). Other CHDs were as follows pulmonary stenosis (7.9%), patent ductus arteriosus (6%), atrioventricular septal defect (3.8%), aortic stenosis (3.5%), dextro-transposition of great arteries (3.5%), tetralogy of fallot (3%), coarctation of aorta (2.8%), and others (26%). Three hundred and ten patients required referral to the cardiac centers in Riyadh for catheterization or surgical intervention. Urgent referral was required in 128 patients. All patients were transferred via medical evacuation service.

Conclusion: Pediatric heart disease constitutes a major health problem in Madina. The majority of patients have CHD. The need for cardiac centre in Madina was emphasized to avoid problems of transportations.

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From the Department of Pediatric Cardiology, Madina Maternity and Children Hospital, Madina, Kingdom of Saudi Arabia.

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Address correspondence and reprint request to: Dr. Abdulhameed A. Alnajjar, Consultant, Pediatric Cardiology Unit, Maternity and Children Hospital, Madina, Kingdom of Saudi Arabia. Tel. +966 (4) 8499616. Fax. +966 (4) 8640936. E-mail: drnajjar2000@yahoo.com

Heart problem in children varies significantly from that in adults. Also pattern of heart diseases in children may vary among different world communities. Heart disease in children remains a major cause of morbidity and mortality. Previous studies demonstrated that congenital heart disease (CHD) constitutes the majority of pediatric heart diseases. Worldwide, CHD are relatively common with a prevalence ranging from 1.0-6.6 per 1000 live births. The prevalence was 1.0 per 1000 in Egyptian children,¹ 2.0 per 1000 in Sudanese children,² 6.6 per 1000 in Americans,³ 5.0 per 1000 in Chinese⁴ and 6.1 per 1000 in Czech Republic.⁵ Although pediatric heart diseases constitute a big health problem, only few studies in Saudi Arabia have been reported. The prevalence of CHD was 2.1 per 1000 in Saudi Arabia.⁶ Knowledge of the epidemiology of pediatric heart diseases is important for assessment of the magnitude of problem, for prevention, diagnosis, management and future plans. There was no previous study to address the magnitude of pediatric heart disease in Madina. The aim of this study is to demonstrate the magnitude of pediatric heart disease problem in Madina, the current situation of pediatric heart service, the obstacles in service and future expectations.

Methods. This was a hospital based, cross sectional study. The study included follow up cases and new cases referred to the Pediatric Cardiology Unit, Madina Maternity and Children Hospital, Madina, Kingdom of Saudi Arabia from January 2007 to June 2008. The work was approved by the ethical committee of the hospital. A computer based software program was established to register all children referred to the pediatric cardiology unit including those who needed further intervention. All referred patients were examined clinically, underwent chest x ray, and electrocardiogram. The diagnosis of all patients was confirmed by echocardiography (Philips Sonos 5500 echocardiography machine (Andover, MA, USA). The result of the echocardiography was confirmed and validated by a team of pediatric cardiology consultants. Each newly diagnosed case was examined by the team together. For sedation, oral chloral hydrate 50 mg/kg body weight was used. The echocardiography examination was conducted using M-mode, colored 2-dimensional, pulse, and continuous wave Doppler echocardiogram. Two-dimensional echocardiographic pictures were recorded in the standard parasternal longaxis, short-axis, apical 4 chamber, subcostal and suprasternal views. The presence and severity of any cardiac defect was analyzed using the American Society of Echocardiography guidelines.⁷ Congenital heart disease were defined as a structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance.⁸ Therefore,

the following conditions were excluded; patent ductus arteriosus (PDA) in premature newborns (one month of age), patent foramen ovale, bicuspid aortic valve without stenosis, persistent left superior vena cava, and right sided aortic arch; and those patients with rheumatic fever without heart involvement. Patients with suspected Kawasaki disease and proven to have another disease were also excluded. Children with functional tricuspid regurg, children with thalassemia and renal failure sent to cardiac function for assessment were also excluded from the study. Patients with sepsis and were sent to rule out vegetation were also excluded. Patients with arrhythmia without structural heart disease were included.

Findings of echo-cardiographic examination were recorded and transferred to SPSS version 12. A descriptive statistical analysis was presented. Patients with structurally and functionally normal hearts were grouped separately. Congenital heart disease was further subdivided into cyanotic and acyanotic heart diseases. Children with cardiomyopathy were placed in the group of myocardial disease. Other disease groups were created. The study was primarily descriptive and included no comparison; therefore, no test of significance was applied.

Results. A total of 4348 children were seen in the study period; 2301 were follow up cases while 2047 were new cases (ranged one day - 14 years). Of the new cases, 794 patients were either congenital or with acquired heart disease while the remaining 1253 children were normal (Figure 1). Of the 794 patients, 705 have CHD (cardiomyopathies 34, rheumatic heart disease 26, mitral valve prolapse 15, Kawasaki disease 9, and arrhythmia 5) (Figure 2).

The most common type of CHD was found to be ventricular septal defect (VSD), and followed by atrial septal defect (ASD) (Table 1).

Table 1 demonstrates the various patients lesions, which were found during the study period. It also demonstrates how many patients needed transfer to cardiac center for further evaluation and/or intervention. Table 2 shows the distribution of major CHD in our study in comparison with other regions in Saudi Arabia including the present study from our hospital. In all regions including Madina, VSD was the most common lesion followed by ASD. Transposition of great arteries was higher in our study than TOF when compared with other regions. No significant difference was found between various regions regarding other defects. Table 3 shows the distribution of major CHD in comparison with studies from Saudi Arabia, Arab countries, and other countries. Of 794 patients, 310 patients needed transfer for a possible or definite further intervention.

This intervention includes either catheter and/or surgical management. Acyanotic CHD constitutes 60% (186 patients) and cyanotic CHD constitutes 33.9% (105 patients). All other lesions constitute 6.1%. All patients were transferred to either one of the 3 cardiac centers in Riyadh.

Discussion. Pediatric heart diseases constitute an important group of diseases that lead to great morbidity and mortality in children. This study does not give a true incidence and prevalence of pediatric heart disease in the total population since it is limited to the Maternity and Children Hospital in Madina. Physicians used to send patients for assessment with very low threshold. In some children, we noticed that echocardiogram was requested even before the chest x-ray. This attitude makes echocardiography laboratory loaded with children, but on the other hand, it protects against passing cardiac patients as normal. Congenital heart disease represented

the majority with a number of 705 patients during the study period and an average of 470 patients per year. This seems to be a large number when compared with those from other regions in Saudi Arabia.⁹⁻¹² When we compared our results with those from the previous study from Madina, it gives a false impression that there was no evidence of any significant increase in the detected new CHD patients.¹³ There is an actual increase in patients because the previous study did not exclude PDA and

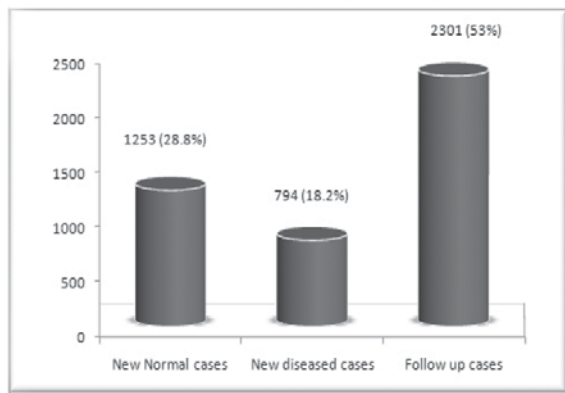


Figure 1 - Distribution of the 4348 children who were examined during the study period.

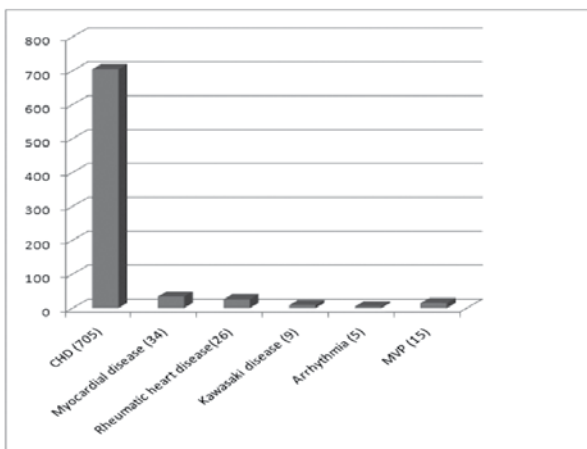


Figure 2 - Pattern of all new lesions during the study period.

Table 1 - Distribution of all new patients heart lesions including those who needed referral to cardiac center for further intervention.

Lesion name	Newly diagnosed cases	Referred to cardiac center n (%)
Atrial septal defect	63	20 (31.7)
Ventricular septal defect	243	28 (11.5)
PDA	42	25 (59.5)
Complete AVSD	22	18 (81.8)
Partial AVSD	5	4 (80.0)
PAPVR	3	3 (100.0)
Mixed left to right shunt	29	17 (58.6)
Mixed acyanotic	70	21 (30.0)
Tetralogy of fallot	21	21 (100.0)
TGA	25	22 (88.0)
DORV	17	17 (100.0)
Pulmonary atresia	10	10 (100.0)
Tricuspid atresia	4	4 (100.0)
HLHS	9	7 (77.8)
Single ventricle	5	5 (100.0)
Isolated V inversion	1	1 (100.0)
Truncus	2	2 (100.0)
Ebstein	1	1 (100.0)
Heterotaxy syndrome	3	3 (100.0)
Absent valve TOF	3	2 (66.7)
Mixed cyanotic	12	7 (58.3)
TAPVR	3	3 (100.0)
Mixed obstructive	7	4 (57.1)
Coarctation of aorta	20	17 (85.0)
Interrupted aortic arch	4	4 (100.0)
Pulmonary stenosis	48	15 (31.3)
Aortic stenosis	25	9 (36.0)
MVP	15	0 (0.0)
BRANCH PS	8	1 (12.5)
DCM	26	2 (7.7)
HCM	8	2 (25.0)
Kawasaki disease	9	2 (22.2)
Rheumatic	26	10 (38.5)
Arrhythmia	5	3 (60.0)
Total	794	310 (39.0)

PDA - patent ductus arteriosus, AVSD - triventricular septal defect, PAPVR - partial anomalous pulmonary venous return, TOF - tetralogy of fallot, TGA - transposition of great arteries, DORV - double outlet right ventricle, HLHS - hypoplastic left heart syndrome, TAPVR - total anomalous pulmonary venous return, MVP - mitral valve prolapse, DCM - dilated cardiomyopathy, HCM - hypertrophic cardiomyopathy,

Table 2 - Frequency of major congenital heart defects in our study compared with that in different regions in Saudi Arabia.

Lesion	Present study	Madina (1997) ¹³	Al Hassa ⁹	Buraidah ¹⁰	Asir ¹¹	Riyadh ¹²
	(1.5 years) n (%)	(3 years) n (%)	(3 years) n (%)	(3 years) n (%)	(2 years) n (%)	(9 years) n (%)
VSD	243 (34.5)	359 (29.7)	292 (39.5)	123 (38.4)	109 (32.5)	127 33.1
ASD	63 (8.9)	314 (26.0)	85 (11.5)	37 (11.6)	35 (10.4)	47 12.2
PS	56 (7.9)	195 (16.1)	66 (8.9)	29 (9.1)	34 (10.1)	20 5.2
PDA	42 (6.0)	159 (13.2)	64 (8.6)	25 (7.8)	53 (15.8)	23 6.0
AVSD	27 (3.8)	38 (3.1)	26 (3.5)	16 (5.0)	12 (3.6)	- -
TOF	21 (3.0)	26 (2.2)	31 (4.2)	15 (4.7)	18 (5.4)	34 8.9
AS	25 (3.5)	20 (1.6)	26 (3.5)	9 (2.8)	9 (2.7)	3 0.8
COA	20 (2.8)	23 (1.9)	20 (2.7)	6 (1.9)	11 (3.3)	17 4.4
TGA	25 (3.5)	22 (1.8)	14 (1.9)	14 (4.4)	5 (1.5)	9 2.3
Others	183 (26.0)	53 (4.4)	116 (15.7)	46 (14.4)	49 (14.6)	104 27.1
Total	705 (100.0)	1209 (100.0)	740 (100.0)	320 (100.0)	355 (100.0)	384 100
Total per year	470	403	246.7	106.7	177.5	42.7

VSD - ventricular septal defect, ASD - atrial septal defect, PS - pulmonary stenosis, PDA - patent ductus arteriosus, AVSD - atrio-ventricular septal defect, TOF - tetralogy of fallot, AS - aortic stenosis, CoA - coarctation of Aorta, TGA - transposition of great arteries.

Table 3 - Distribution of major congenital heart defects in our study in comparison to total Saudi Arabia and international studies.

Country/City	VSD	ASD	PDA	TOF	AVSD	AS	PS	COA	TGA	Others
Present study	34.5	8.9	6.0	3.0	3.8	3.5	7.9	2.8	3.5	26.0
Saudi Arabia (2006) ¹⁴	33.9	18.1	11.6	3.5	3.5	2.5	12.4	2.3	2.1	10.1
Oman (2000) ¹⁵	24.9	14.4	10.3	9.6	5.9	3.6	8.8	3.7	3.6	15.2
Qatar (1997) ¹⁶	40.6	7.2	5.1	5.1	2.8	2.5	8.7	4.1	3.1	20.8
Kuwait (2004) ¹⁷	33.2	9.7	19.0	4.1	7.6	2.1	7.6	4.8	3.3	21.6
Egypt (2000) ¹	35.3	13.6	6.1	5.0	3.1	3.9	18.4	0.9	2.0	11.7
Jordan (2008) ¹⁸	43.4	13.6	8.3	9.5	3.6	4.3	6.2	3.4	5.5	2.2
Sudan (2007) ¹⁹	16.1	6.2	4.6	17.7	8.6	1.3	6	0.7	6.6	32.2
Lebanon (1999) ²⁰	31.0	9.0	6.0	8.0	-	4.0	15	2.0	3.0	22
Iran (2008) ²¹	11.1	19.5	18.0	17.0	0.6	0.0	3.6	1.7	0.1	28.5
Turkey (2006) ²²	32.6	13.1	15.9	5.8	3.6	4.5	7.9	4.8	3.0	8.8
Nigeria (2007) ²³	45.9	12.3	2.5	26.0	8.2	-	-	-	-	4.9
India (2006) ²⁴	30.5	14.4	7.2	10.0	0.1	0.3	0.3	-	0.6	36.6
UK (1981) ²⁵	32.5	5.9	11.9	5.9	2.4	5.1	7.6	6.3	5.0	17.4
USA (1990) ³	32.1	7.4	8.3	3.8	3.6	3.8	8.6	6.7	2.6	23.1
USA (2008) ²⁶	41.8	13.1	2.9	4.7	4.1	1.1	5.5	4.4	2.3	20.1
Bohemia (1999) ⁵	41.6	8.7	5.1	3.4	4.0	7.8	5.8	5.3	5.4	12.1
Denmark (1980) ²⁷	24.0	9.4	12.6	5.8	2.6	4.7	5.9	7.0	4.8	23.2
Australia (2004) ²⁸	57.4	11.1	7.4	1.9	0.9	1.9	1.9	1.9	0.9	14.8
Japan (1990) ²⁹	60.0	5.3	3.6	5.8	1.8	1.0	9.6	2.7	2.2	9.5
China (2001) ³⁰	54.0	9.2	7.9	5.3	2.6	2.6	2.6	1.3	4.0	10.5

VSD - ventricular septal defect, ASD - atrial septal defect, PS - pulmonary stenosis, PDA - patent ductus arteriosus, AVSD - atrio-ventricular septal defect, TOF - tetralogy of fallot, AS - aortic stenosis, CoA - coarctation of aorta, TGA - transposition of great arteries.

ASD during the neonatal period which constituted a bulk of patients (138 patients during the study period). This is also the reason for the big difference of numbers of the 2 defects between the 2 studies. The frequency of the major CHD in our study is consistent with that of total Saudi Arabia distribution.¹⁴ Ventricular septal defect, is the most common lesion worldwide except in studies from Sudan, Nigeria (TOF), and Iran (ASD). Ventricular septal defect ranges from 11.1-60% in various studies in Iran (the lowest) and Japan (the highest). On the other hand, ASD distribution had a reverse pattern with highest value in Iran and lowest value in Japan.¹⁵⁻³⁰

Rheumatic heart disease is the most common acquired heart lesion. In countries where efficient treatment of streptococcal infection was achieved, KD replaced RHD as the most common acquired heart disease. This indicates that we still in need to pay more effort to prevent occurrence of rheumatic fever in our children. Parental education on the serious effects of incompletely treated or neglected streptococcal infection should be carried out. Compliance to regular long acting penicillin, administration should be encouraged. In patients with KD, coronary lesion was detected in 3 patients. Delay in diagnosis and delay in immunoglobulin therapy is a risk factor for developing coronary artery lesion. Awareness of the KD criteria especially for physicians in the periphery area may help to decrease such problems in the future. Cardiomyopathies represent a considerable portion of the heart problem in our children. We found that some patients follow viral myocarditis and most of them shows good prognosis. Other patients have poor prognosis and they are either sporadic cases or familial cases. More detailed study including genetic analysis is required in such patients especially those with familial tendency. Ventricular septal defect represents the most common CHD in our study. Worldwide, VSD is the most common CHD with few exceptional studies. The percentage of VSD varies among different countries with a range from 8-60%.¹⁵⁻³⁰ The distribution of various single CHD varies from region to another and from country to another. This is may be explained by the difference in genetics and ethnicity. In our study and previous studies from other regions in Saudi Arabia, ASD constitutes approximately 10% of CHDs.⁹⁻¹² A previous combined studies from different regions in Saudi Arabia concluded a percentage of ASD to be 18.1%.¹⁴ We observed that this percentage was too high because they included ASDs in neonates which resulted in bias impression. The percentage of ASD varies from 5.3-19.5%.¹⁵⁻³⁰ In patients with Down's syndrome, AVSD is the most common results of CHD. In our study, AVSD lesions were found in patients with Down's syndrome with

some exception (2 patients) due to heterotaxia. Dextro-transposition of great arteries (D-TGA) in our study was more than TOF. Other studies have TOF percentage which is more than the D-TGA except study in the study of Bohemia.⁵ It is well known that in the neonatal period, we find that the D-TGA is the most common cyanotic CHD due to early presentation. The TOF was unnoticed during this period. In our hospital, we have a busy neonatal unit with more than 80 beds. This may be the cause of having more D-TGA patients than TOF. The percentages of other CHDs go in agreement with those from other areas in the world with variations which can be attributed to ethnic and genetic factors. Approximately 40% of our patients needed transfer to either one of the 3 cardiac centers in Riyadh for a possible catheter and/or surgical intervention. The catheter- or surgical- based intervention are not yet available in Madina. The vast majority of patients transferred to the cardiac centers were suffering from CHDs. And we found more cyanotic CHD that required further intervention in our study. But few of them need not to transfer due to inoperability of complex lesions, other major associated anomalies major chromosomal abnormality, or death of patients.

Critically ill patients are frequently transferred using the air ambulance service called medical evacuation (MEDEVAC). Some of the problems we faced during the transfer are the unstable patients.

Another problem that we encountered is the shortage of bed in the cardiac center especially in the pediatric intensive care unit (PICU). This shortage may lead to increased morbidity and mortality. According to the recommendations of the American Academy of Pediatrics, a pediatric cardiac center should be available for a population that generates more than 30 000 live births per year.³¹ Applying the above recommendation, a cardiac center with full facilities is now under construction and this will be a great service to the region.

In conclusion, pediatric heart disease constitutes a major health problem in Madina. Rheumatic heart disease is still the major acquired heart disease and needs more parental education. About 40% of patients with pediatric heart diseases needed referral to cardiac center. Thus, the need for cardiac center in Madina was emphasized to avoid problems of transportations.

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