

## Congenital heart disease in south-west Saudi Arabia

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Congenital heart disease (CHD) constitutes the majority of pediatric heart diseases and remains one of the major causes of morbidity and mortality. Previous studies demonstrated that CHD worldwide, is relatively common with a prevalence ranging from 4-50 per 1000 live births.<sup>1</sup> The prevalence was 10.7 per 1000 in KSA<sup>2</sup> in 2001, and 17.5 per 1000 in Central Australia<sup>3</sup> in 2004. Only a few studies in Saudi Arabia have been reported. The prevalence of CHD varies from study to study due to trivial defects included or excluded. Knowledge of the epidemiology of pediatric heart diseases is important for assessment of the magnitude of the problem, for prevention, diagnosis, management, and future plans. There is limited information on the magnitude of the pediatric heart disease problem in Albaha, Kingdom of Saudi Arabia. The aim of this study is to demonstrate the magnitude of CHD in Albaha, the current situation of the pediatric heart disease service, the obstacles in the service, and future expectations.

This is hospital based, cross sectional study, included follow up and new cases referred to the Pediatric Cardiology Unit, King Fahad Hospital Albaha, Kingdom of Saudi Arabia from April 2005 to 2010. All patients were aged between birth and 12 years old. The work was approved by the ethical committee of the hospital. A computer based software program was established to register all children screened in the study. All referred patients were examined clinically, underwent chest x-ray, and electrocardiogram. The diagnosis of all patients was confirmed by echocardiography (Philips IE33 echocardiography machine, Philips, Bothel, WA, USA). 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 long axis, short axis, apical 4 chamber, subcostal and suprasternal views. The presence and severity of any cardiac defect were analyzed using the American Society of Echocardiography guidelines. Congenital heart disease was defined as a structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance. 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, trivial mitral regurgitation (MR), trivial tricuspid regurgitation (TR). Patients with suspected Kawasaki disease without cardiac impairment, children with thalassemia and other hematology problems, rheumatic heart disease, and other acquired heart diseases were also excluded. Patients with cardiac murmur with structurally and functionally normal hearts were excluded and grouped separately. Congenital heart disease was subdivided into cyanotic and acyanotic heart diseases.

A total of 2610 children were screened in the study, 400 patients as follow up cases diagnosed before 2005, while 2210 children were new cases screened within the study period in our pediatric cardiology unit due to cardiac murmur by clinical examination. Five hundred and thirty children had normal echocardiography, 500 cases were preterm with PDA, and 480 cases were with patent foramen ovale. Seven hundred patients (31.8%) of these new cases were diagnosed with heart disease, 591 (26.8%) of them were found to have CHD. The most frequent type of CHD are shown in Table 1 and were, double outlet right ventricle in 10 patients (1.7%), pulmonary regurgitation 7

**Table 1 -** Frequency of main types of congenital heart diseases in our study compared with other regions in Kingdom of Saudi Arabia.

CHD	Albaha 2010	Madina <sup>6</sup>	Alhassa <sup>8</sup>	Buraidah <sup>9</sup>	Asir <sup>10</sup>	Riyadh <sup>7</sup>
n (%)						
VSD	175(29.6)	243(34.5)	292(39.5)	123(38.4)	109(32.5)	127(33.1)
PDA	56 (9.5)	42 (6.0)	64 (8.6)	25 (7.8)	53(15.8)	23 (6.0)
ASD	55 (9.3)	63 (8.9)	85(11.5)	37 (11.6)	35(10.4)	47(12.2)
PS	47 (7.9)	56 (7.9)	66 (8.9)	29 (9.1)	34(10.1)	20 (5.2)
AVSD	36 (6.0)	27 (3.8)	26 (3.5)	16 (5.0)	12 (3.6)	--
TOF	27 (4.7)	21 (3.0)	31 (4.2)	15 (4.7)	18 (5.4)	34 (8.9)
COA	20 (3.4)	20 (2.8)	20 (2.7)	6 (1.9)	11 (3.3)	17 (4.4)
AS	18 (3.0)	25 (3.5)	26 (3.5)	9 (2.8)	9 (2.7)	3 (0.8)
TGA	11 (1.9)	25 (3.5)	14 (1.9)	14 (4.4)	5 (1.5)	9 (2.3)

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

patients (1.2%), other CHD included pulmonary atresia 7 patients (1.2%), truncus arteriosus 7 patients (1.2%), hypoplastic left heart syndrome 7 patients (1.2%), total anomalous of pulmonary venous returns, 6 patients (1%), infant of diabetic mother with left ventricular outflow tract obstruction 5 patients (0.8%), dextrocardia (mirror image) 5 patients (0.8%), congenital mitral stenosis 3 patients (0.5%), aortic regurgitation, 2 patients (0.3%), arrhythmogenic right ventricular hypertrophy, one patient (0.2%), libman-sacks endocarditis, one patient (0.2%), and other congenital defects 38 patients (6.4%). A total of 523 patients (88.5%) from all patients with CHD were diagnosed as acyanotic CHD, while 68 patients (11.5%) as cyanotic CHD.

This study does not give a true incidence and prevalence of CHD in the total population since it is limited to the King Fahad Hospital at Albaha. Congenital heart diseases represented the majority with a number of 591 patients during the study period, and an average of 120 patients per year. This seems to be a large number when compared with the total population of this area. When we compared our results with those from other regions of Kingdom of Saudi Arabia. It gives an impression that CHDs in Albaha area in the same range, with a higher percentage of atrioventricular septal defect especially in children with Down's syndrome (Table 1). Also, the reason for the difference from one study to another may be due to trivial defects, which were included in some studies and excluded in others. The frequency of the major types of CHDs in our study is consistent with that of the total Saudi Arabia distribution. The VSD is the most common lesion worldwide, and represents the most common defect, it was 29.6% in our study; as in all international studies, which range between 24 to 60% with a few exceptional studies, it is 41.8% in USA.<sup>4</sup> On the other hand, ASD is 9.3% in our study and 18.1% in other study from the Kingdom of Saudi Arabia.<sup>5</sup> This high percentage in other studies from other regions in the Kingdom (Table 1) may be because they included ASDs in neonates, which resulted in bias impression. Atrioventricular septal defect represents 36 patients 6% in our study, which is the highest percentage in Kingdom of Saudi Arabia regions (Table 1). Thirty patients of them (83.3%) had down's syndrome. This ranges from 0.1-6.7% in international studies.

The Tetralogy of Fallot percentage (4.7%) is higher than d-Transposition of great arteries percentage (1.9%) in our study like in other studies. We found that the percentages of other types of CHD in our study were similar to other worldwide studies. The male to female ratio in our patients is 47.3% in males to 52.8% in females. Approximately 40% of our patients needed transfer to one of the higher cardiac centers in Riyadh or Jeddah for further evaluation or interventional cardiac catheterization or surgical correction. 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 beds in the cardiac center, especially in the, Pediatric Intensive Care Unit. This shortage may lead to increased morbidity and mortality. According to this high percentage of CHD, a cardiac center, with full facilities is now under construction and this will be a great service to this region.

In conclusion, CHD constitutes a major health problem in Albaha. Approximately 40% of our patients with CHDs needed referral to higher cardiac centers. An increase in atrioventricular septal defect cases was observed mostly seen in patients with Down's syndrome (trisomy 21) Health education is recommended for prevention of this syndrome. Detailed study including genetic analysis is required in such patients especially those with familial tendency of CHD.

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