Congenital heart disease profile: Four perspectives

This issue of annals of pediatric cardiology has four large studies that describe the profiles of congenital heart disease (CHD) in four different situations from India and Pakistan.

The first study[1] describes CHD profile at birth in a large community hospital in North India. The second study presents a CHD profile in a tertiary care national referral hospital in Pakistan with facilities for infant heart surgery.[2] This study seeks to examine the delay in referral to a tertiary care center. The third study describes the CHD profile in a medical college hospital in North India that does not have facilities for infant heart surgery and seeks to identify risk factors for CHD using a case–control design.[3] Finally, the fourth study describes the result of surgery for CHD among adults referred to an advanced tertiary referral hospital in North India.[4] This study is included in this editorial because it offers a window to understand the profile of adults, most of whom had untreated or previously palliated CHD.

These four studies offer a unique opportunity to understand the potential factors that influence CHD profile in different age groups in low- and middle-income nations, wherein the vast majority of children born with CHD do not receive timely attention.

The first study by Saxena *et al.*[1] is one of the largest studies involving newborns from low- and middle-income countries (LMICs). The strength of the study was the fact that every single newborn underwent an echocardiogram. The profile at birth, therefore, is the most accurate available datum from LMICs. This profile appears similar to the previously published studies from advanced nations, with ventricular septal defects (VSDs) emerging as the most common heart defect. Nearly, one-fifth of the newborns with a significant CHD had cyanotic CHD, and transposition and hypoplastic left heart syndrome (HLHS) were the most common. The frequency of Tetralogy of Fallot (TOF) was lower and similar to a number of other cyanotic heart defects that were identified.

The second study from Pakistan[2] is a cross-sectional study that looks at 354 consecutive children under the age of 15 years who were referred during a 6-month period to the only pediatric heart program with the capability of infant heart surgery serving a population of 100 million. Apparently, these numbers only represent a small proportion of all children with CHD in the region. A median age of 2 years suggests that most newborns and infants with CHD never received timely attention. Indeed, most were delivered at home or in rural maternity centers, largely limiting the possibility of early diagnosis of CHD. A male–female ratio of 1.7:1 is also compatible with a strong gender bias among families seeking care for their children. The high prevalence of severe undernourishment reflected the socioeconomic status of the families as well as the impact of CHD on nutritional status. It is reasonable to assume that the CHD profile in the referred population would represent a population selected by natural history. The small proportion of patients with hemodynamically insignificant defects (such as a restrictive VSD or patent ductus arteriosus) and a strikingly small proportion of children with atrial septal defect (ASD) perhaps reflect the fact that families of children who do not have symptoms are unlikely to seek attention. Not surprisingly, therefore, the large VSD is by far the most common heart defect in this study population. Among the cyanotic defects, TOF emerges as the most common lesion, clearly underscoring its relatively favorable natural history. HLHS does not figure in the list of conditions suggesting that all patients with HLHS die early. The number of patients (16 patients) with transposition of the great artery (TGA) and intact ventricular septum (IVS) is larger than what one would have expected. The median delay in the diagnosis of 4 months for TGA-IVS is not surprising. Another conclusion that can be drawn from this statistic is the fact that for every TGA-IVS that reaches the hospital in these circumstances, there are likely to be several that die in the newborn period.

The third study from Aligarh,[3] North India, was from a government medical college. This center had limited capability to provide comprehensive pediatric heart care and did not offer infant or newborn heart surgery. All CHD patients clinically identified in the department of pediatrics

Access this article online

Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/0974-2069.189110

How to cite this article: Kumar RK. Congenital heart disease profile: Four perspectives. Ann Pediatr Card 2016;9:203-4.

and subsequently confirmed through echocardiography were included. A gender bias favoring male children was seen in this study here as well. The profile has some similarities when compared to the study from Pakistan. Here again, TOF emerged as the most common cyanotic CHD, TGA was rare, and HLHS was nonexistent. The differences may reflect the fact that there was a greater opportunity to identify relatively asymptomatic patients with CHD through clinical examination. As a result, there are a larger number of patients with ASD in this study when compared to the study from Pakistan.

Finally, in the study from the All India Institute of Medical Sciences, New Delhi, Talwar *et al.* present a large experience with CHD surgery among adolescents (>13 years age) and adults involving 1142 patients over a 10-year period.[4] While the primary purpose of the study was to describe risk factors for adverse surgical outcomes, this editorial's focus will be on the profile of the patients who underwent surgery. As expected, the vast majority of the patients were not previously operated, and the single largest lesion category was ASD. Among the cyanotic conditions, TOF was by far the most common. Among the 314 patients undergoing TOF repair, 262 (83%) did not require a transannular patch suggesting that they were at the less severe end of the spectrum of the right ventricular outflow tract spectrum, again underscoring the impact of natural history on the patient profile.

It is clear from these studies that the profile of patients with CHD that present to health-care facilities in LMICs is largely determined by the natural history of individual conditions. A high attrition of CHD patients with conditions that are known to have a high early mortality results in low frequency of these lesions encountered in many settings in LMICs and may contribute to the prevailing perceptions on their rarity. Other significant determinants include robustness of health systems and the quality of maternal and child health services. Most newborns with critical CHD are likely to be missed altogether when they are delivered at home or in poorly equipped facilities. In addition, limited awareness on CHD diagnosis among pediatricians who staff larger facilities is also likely to contribute to CHDs being missed. The comparison of profiles in the studies presented here clearly suggests that majority of the newborns with correctable CHDs such as TGA and total anomalous pulmonary venous connection are dying without a diagnosis.

The results of these studies have important public health implications as follows:

1. To make meaningful reductions in mortality and morbidity from CHD, it is imperative to focus on comprehensive newborn and infant cardiac care. The institutions that seek to develop infant and newborn cardiac services need to be affordable to the average family in the region
2. It is somewhat futile to focus on developing advanced pediatric heart programs without parallel improvements in maternal and child health services. A number of elements need to be looked into. These include substantial improvement in the proportion of institutional deliveries, improved quality of perinatal care in primary care settings, improving awareness and training of primary care professionals on infant and newborn heart disease detection, and carefully considered implementation of newborn screening for congenital heart defects[5]
3. Transport of newborns and infants with CHD is a seriously neglected issue in India and other LMICs. There is practically no organized system for safe transport of newborns and infants with CHD in most LMICs. The average distances traveled by families are substantial. The risks of hypothermia, hypoglycemia, sepsis, and electrolyte imbalance add substantially to the already serious situation resulting from CHD during the several hours spent in largely unsupervised transport.

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