INCIDENCE OF CONGENITAL HEART DISEASE AMONG HOSPITAL LIVE BIRTHS IN INDIA

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ABSTRACT

Ten thousand nine hundred and sixty four consecutive live births weighing more than 500 g and more than 28 weeks of gestation were subjected to a thorough clinical examination within 24 h of birth. Those suspected of having congenital heart disease (CHD) were followed up every 4-6 weeks for a period of 6 to 18 months (mean 9.75 months).

Forty three of 10,964 infants had CHD, i.e., 3.9/1000 live births. Incidence of CHD was higher in pre-terms as compared to full term live births (22.69 vs 2.36/1000 live births). Diagnosis was confirmed by echocardiography including 2D, Doppler and color flow imaging. Twenty eight per cent of the infants with CHD had other associated somatic anomalies, Down's syndrome being the commonest (9.3%). Patent ductus arteriosus (41.9%) and ventricular septal defects (VSD) (34.9%), were the commonest lesions with an incidence of 1.6 and 1.4/1000 live births, respectively. Incidence of PDA was higher probably because of larger number of pre-term deliveries. During follow up of 6-18 months, 34.9% of the infants with CHD died. The diagnosis of CHD was confirmed at autopsy in 20% of the deaths.

Key words: *Congenital heart disease, Prematurity, Mortality.*

The incidence of congenital heart disease (CHD) depends upon various factors—the nature of the sample (all live births or all births), the source of information (birth or death certificates) or on the spot examination by a pediatric cardiologist. The incidence is higher if early spontaneous abortions are also included. Recent studies have reported incidence of CHD to range between 4.05 to 10.2/1000 live births. Earlier reported incidence of CHD of 3-5/1000 live births was low because of inadequate diagnostic techniques and availability of ineffective surgical therapy(l).

This prospective study was conducted to ascertain the incidence of CHD in live births with subsequent confirmation of diagnosis by echocardiography and color flow mapping and also to study any difference in the incidence of CHD in preterm and term live births.

Material and Methods

All ten thousand nine hundred and sixty-four live newborns delivered at Lok Nayak Jai Prakash Narain Hospital, New Delhi, India during 28 months constituted the material of the study. These newborns were more than 500 grams and were products of more than 28 weeks gestation. A thorough clinical examination was carried out within 24 hours of birth and CHD was suspected in presence of the following criteria as

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defined by Mitchell *et al.(2):* presence of a cardiac murmur, presence of cyanosis or feeding difficulty only, or cyanosis associated with feeding difficulty. Presence of features of congestive heart failure or failure to thrive, and detailed information was collected with special reference to family history of congenital heart disease in siblings, parents and first degree relatives. Significant antenatal history, for example, history of irradiation or drug or hormonal ingestion, exanthematous fever in 6 months prior to conception, first trimester bleeding or antepartum hemorrhage, maternal age and parity and information on baby's birth weight, sex and gestational age were elicited. All suspected patients were investigated with roentgenograms, electrocardiograms, and were followed up for a period of 6-18 months (mean 9.75 months) at 4-6 weeks interval. Echocardiographic examination was performed on Diasonics, cardio-imaging phased array ultrasonograph with an inbuilt m-Mode, 2D, black and white color doppler. Standard supine with head extended and left lateral positions were used to achieve parasternal, apical, subcostal and suprasternal views. Autopsies were performed whenever it was possible within 6 h of death.

Abnormalities of the systemic veins such as persistent left superior vena cava or vena cava—azygous continuity, abnormalities of systemic artery branches such as combined innominate—left carotid arterial trunk and arrhythmias associated with structural malformation were excluded. Patent ductus arteriosus in the first 14 days of life without causing CHF was considered normal. Various murmurs were classified as follows:

Transient Murmurs: Murmurs not associated with structural cardiac malfunction during detailed clinical examination and echocardiographic evaluation and which disappeared on subsequent examination.

Innocent Systolic Murmur: Murmur in which cardiological examination and echocardiography ruled out structural cardiac malformation.

Definite CHD: A diagnosis of definite CHD was based on obvious clinical findings of CHD as confirmed by a pediatric cardiologist, echocardiographic data or on autopsy findings.

Results

During 28 months, 11,213 deliveries were conducted, of which 10,964 (92.2%) were live births and 249 (7.8%) were still births; 10,125 (92.2%) were full term and 839 (7.8%) were preterm deliveries, (those who were born before completing 37 weeks gestation), 30.8% were less than 2500 g and 69.2% were more than 2500 g (full term babies).

Incidence of CHD

Ninety six babies were suspected to have congenital heart disease on first examination. Of these 43 (44.8%) had definite CHD. Innocent murmur was detected in 29.2% babies *(Table I).* The age for disappearance of transient murmurs ranged from one to 3 months (mean 20.1 ± 10.1 days).

Birth Weight and Gestation Period

Of 43 babies (3.9/1000 live births), 22 were males and 21 females. The incidence of congenital heart disease in preterm babies was significantly higher than term babies (22.86/1000 *vs* 2.36/1000) ($p<0.001$) *(Table II).*

Clinical Presentation

On the first day, cyanosis was detected in 5 (11.6%) of CHD cases. Twelve (27.9%)

TABLE I—*Outcome of Suspected Cases of Hearth Diseases*

Outcome	No. of cases	Percentage of suspected cases	Incidence per thousand live births
Definite CHD	43	44.8	3.92
Transient murmurs	25	26.0	2.28
Innocent murmurs	28	29.2	2.50

TABLE II—*Distribution of CHD by Birth Weight and Gestation*

Gestational age (wks)	No. of children (No./1000 live births)	Birth weight (g)		
		<1500	1500-2500	>2500
<37	19 (22.86)	13	6	–
>37	24 (2.36)	–	15	9
Total	43	13	21	9

and 7 (16.2%) babies presented with murmur and cyanosis, respectively in the first week. The incidence of murmur being detected beyond one week increased to 34.9% of CHD cases. The earliest murmur could be detected after 24 hours of birth and it was ejection systolic in nature. Congestive heart failure was another important manifestation of CHD seen in 11.6% beyond one week.

Somatic Malformations and CHD

Somatic anomalies were associated in 17.9% of patients with CHD. Down's syndrome was the commonest anomaly (9.3%, n=4) followed by congenital talipes equinovarus (6.4%, n = 3), anencephaly (4.6%, n = 2) and craniosynostosis and micrognathia accounted for one case each (2.3%).

Confirmation of CHD

Clinical diagnosis was confirmed in 30 babies (70%) by echocardiography, during surgery in 2 (4.65%) and by autopsy in 3 (6.9%). On echocardiography, VSD and PDA were the commonest lesions found in 34.8% and 18.6% respectively *(Table III)*. The rest of the patients were too sick to be taken for echocardiographic examination and they expired subsequently.

Mortality in CHD

Mortality of CHD in the present series was 23.2% (10 out of 43). Four of the 10 babies (40%) died within 72 hours of birth and all of them were cyanotic heart disease. One of them had tricuspid atresia and was successfully subjected to atrial septostomy. Two children died of refractory cardiac failure. Two of these 4 babies were subjected to autopsy and had complex congenital heart anomalies. Two babies of PDA died between 3-7 days of refractory congestive heart failure. Prematurity complicated by

TABLE III–*Echocardiographic Confirmation of CHD (n=30)*

S.No.	Lesion	No.	Percentage
1	VSD	15	34.8
2	PDA	8	18.6
3	ASD	1	2.3
4	TOF	2	4.6
5	Valvular AS	1	2.3
6	Endocardial fibroelastosis	1	2.3
7	Tricuspid atresia	1	2.3
8	Single Ventricle with TGA	1	2.3

Total cases were 43.

hyaline membrane disease and CHF was the cause of death in this group. Another 40% (n = 4, 3 with VSD and 1 with PDA) babies died after 7 days. Autopsy performed in 3 cases confirmed single atrium, single ventricle with truncus arteriosus in 2 and complete A-V canal with large subaortic VSD (20 mm) in the third case. *Figures 1-3* show the echocardiographic and autopsy findings of some of the cases with CHD.

Discussion

Congenital heart disease (CHD) has become increasingly important in recent years because it is one of the important causes of morbidity and mortality in infancy and also there has been a decline in rheumatic fever in affluent societies. CHD along with neural tube defects account for two-thirds of all major birth defects(3).

The reported incidence of CHD in live new borns tends to vary a lot due to various unrecognizable lesions at birth and lack of technical expertise(4). The reported incidence in the Western literature are 0.65/1000(5), 1.8/1000(6), 8.3/1000(7), 9/1000(8) and 11.9/1000(9). Indian studies incidence varies from 1.45/1000(10) to 4/1000(11).

Ferencz *et al.*(12) reviewed seven major studies from Europe and North America and concluded that confirmed CHD incidence had been remarkably constant at 4/1000 live births over 40 years time span from 1940 to 1980. The incidence reported in the present study of 3.9/1000 live birth is in close proximity.

The variable rate of fall of pulmonary vascular resistance (PVR) after birth and variable time sequence for closure of PDA result in variable rate of detection of CHD at different time intervals. Most workers agree that 50% of all cases of CHD are detected by 1 month of age, 75% by 3 months and 100% by 3-4 years of age (4,12-14). It is, therefore, suggested that ideal study should follow all live births upto 5 years of age. Data from nurseries without adequate follow up gives a very low incidence(6,10). In the present study, 35.3% were detected within 72 hours of birth, 70.6% by one month and remaining by 3

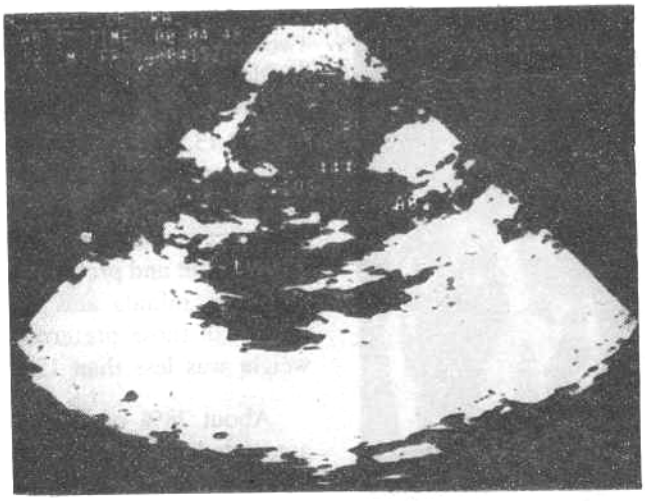

Fig. 1. PSLA view showing large subaortic VSD and over-riding of aorta in tetralogy of Fallot.

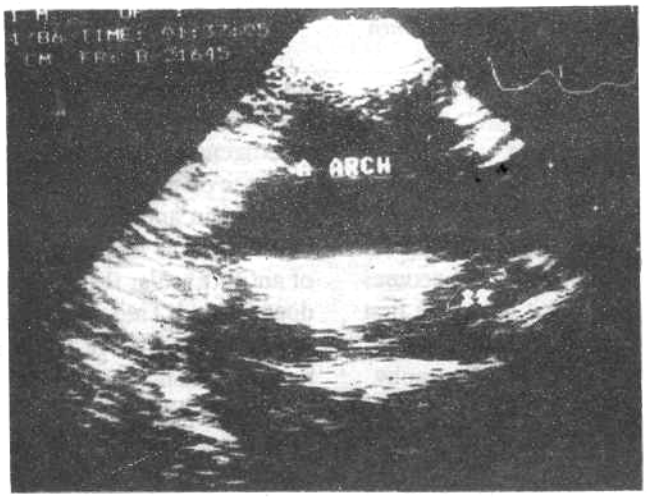

Fig 2. 2DE showing PDA in SSLA view.

months. Keeping in mind the above time frame of CHD detection, it is expected that about another 25% could have been detected beyond 3 months of age which may give still higher incidence of CHD in our setting.

The diagnosis of CHD is fallacious on clinical grounds alone, the variable incidence reported in literature especially in Indian studies(15-18) were due to lack of supplementation of echocardiography. Ferencz

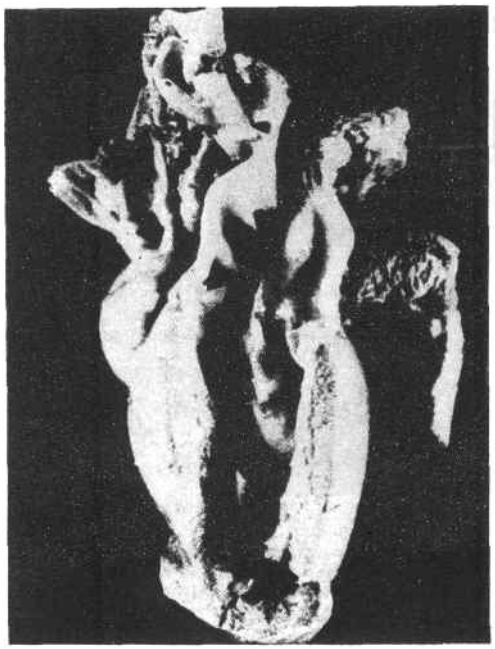

Fig 3. Complex heart disease—autopsy specimen showing univentricular heart

etal.(12) reported CHD incidence of 3.97/1000 live births when supplemented by echocardiography which was similar (3.9/1000) to that reported in this study. It becomes significant in the light that it being the first Indian study where 70% confirmation of diagnosis was done by echocardiography.

Most studies(3,17) have reported equal incidence of CHD in either sex as also reported in this series. There has been no seasonal variation in the incidence of any particular type of CHD in our study in contrast to reports of differing seasonal peaks observed specially for PDA(4,18,19). The age range of mothers 17-32 years and of fathers 27-38 years were equally distributed in all groups of CHD babies.

Birth weight and period of gestation are two important determinants of CHD, the incidence is reported to be 2-3 times higher in low birth weight and preterm infants(3,13,20). Yerushalmy(13) reported higher incidence of CHD in low birth weight full term infants. In the present series the incidence was about 10 times higher in low birth weight and preterms in comparison to full term infants and the incidence was highest in those preterms where the birth weight was less than 1500 g.

About 28% of infants with CHD had associated congenital somatic anomalies. The incidence of Down's syndrome is 1.5/1000 live births(21) and in association with CHD varies from 11.7% to as high as 40% (22,23). The incidence of CHD in association with Down's syndrome was 9.3% in the present series and appears to be independent of maternal age(24).

Various drugs, *e.g.,* phenytoin, amphetamine, coumarin(24), alcohol, smoking, thalidomide(25) have all been implicated in teratogenesis. In this series only two mothers were on antitubercular treatment (INH, Rifampkin and Ethambutol) for 6 months and newborns had normal heart. The role of antitubercular therapy in CHD is not yet documented. There is always concern about the increased risk of CHD among the siblings of patients with CHD(17,26-29). Nora and Nora(28) identified high risk families where there is higher incidence of CHD in the first degree relatives. To our surprise none of the newborns had siblings or first degree relatives suffering or having a history of CHD (history of murmur in infancy).

The reported frequency of individual cardiac lesions depends upon the diagnostic methods used and duration of follow up

(since a small VSD may close). VSD is the commonest lesion (27-32%) reported, followed by PDA (2.6-11.9%). The other common lesions are ASD, PS coarctation of aorta *(Tables TV and V)*. The present series document PDA (41.9%) and VSD

TABLE IV–*Percentage Distribution of Types of CHD in Live Born Infants*

Lesion	Gothenburg	USA Multi-centre	Black Pool	California	Liverpool	Baltimore Washington
Reference	(27)	(24)	(4)	(29)	(14)	(12)
VSD	27.1	32.1	28.1	31.3	32.5	26.3
PDA	9.5	8.3	6.5	5.5	11.9	2.6
ASD	4.3	7.4	8.3	6.1	5.9	7.5
PS	3.8	8.6	2.7	13.5	7.6	7.0
AS	5.4	3.8	4.1	3.7	5.1	3.3
Coarctation	9.8	6.7	5.6	5.5	6.3	6.8
d TGA	6.0	2.6	5.6	3.7	5.0	5.0
TOF	4.1	3.8	8.6	3.7	5.9	9.2
TA	1.4	1.7	1.2	2.5	1.1	1.5
HLH	0.8	3.1	3.3	0.6	2.8	5.7
SV	0.0	0.0	1.5	0.6	1.7	0.0
TAPVC	0.8	0.0	2.1	0.6	0.8	1.7
Misc	21.7	13.8	13.6	17.8	8.0	11.6

TABLE V–*Classification of CHD (n=43)*

Total	No	%	Per 1000 live births
VSD	15	34.9	1.3
with ASD	1		
with PDA	1		
PDA	18	42.0	1.6
ASD	1	2.3	0.1
TOF	2	4.6	0.2
T. atresia	1	2.3	0.1
*EFE	1	2.3	0.1
**A.S.	1	2.3	0.1
+SVT TGA	1	2.3	0.1
Complex cyanotic anomaly	3	7.0	0.3
	43	100.0	3.9

* Endocardial fibroelastosis.
** Aortic stenosis (valvular).
\+ Single ventricle with transposition of great arteries.

(34.9%) as the commonest lesions giving an incidence of 1.6/1000 and 1.3/1000 live births, respectively. The incidence of PDA was higher in the present series in comparison to the quoted studies probably because of increased number of preterm deliveries in our settings.

The neonatal mortality besides being influenced by available facilities for early detection, supporting care and surgical correction is also dependent upon the level of perinatal care because of prematurity, low birth weight, asphyxia, infections, *etc.* These factors are responsible for 60% of neonatal deaths among infants with CHD(27). The neonatal mortality in the present series accounted for 23.25% deaths, which was similar to that reported by Carlgren(27) 22% and slightly higher than that reported by Yerushalmy(13) *i.e.,* 15%.

Thus, apart from the size of sample studied, to find out the incidence of CHD among live births, more emphasis lies on comprehensive examination of newborns at frequent intervals and the modes of diagnostic techniques.

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