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The present study describes an effort to have a surveillance for congenital heart diseases. This study has been carried out on patients of various types of congenital heart diseases, who attended the out-patient department of Pediatrics or admitted in Pediatric ward of M.L.B. Medical College Hospital, Jhansi from September 1989 to August, 1990.

As per outdoor patient records of Pediatrics department, the prevalence of congenital heart disease was found to be 1.45%. Higher prevalence was reported by Padmavati and Datey (1968) from various Indian city hospitals as 4.8% in Delhi (1952-1956), 2.3% in Madras (1946), 3.6% in Amritsar (1953), 6.3% in Bombay (1952-1956) and 1.6% in Lucknow (1953). Banerjee et al found it to be 7.5% in all autopsy study from 1964-1975 at P.C.I., Chandigarh. Such data were mostly from large referral hospitals, so there, it is likely to be some concentration of cardiac cases. Moreover, their figures were of patients of all age groups both pediatric as well as adults. Nevertheless, we admit some dilution of data in our study due to undiagnosed cases in the register. Various other studies are also available from different countries, where incidence of congenital heart disease was calculated at the
time of birth varied from 0.7% to 1.17% of live births as reported by Neel (Japan) 1948-1954, Richards et al (New York) 1946-1953, Kerebijin (Holland) 1958, Yerushalmy (California) 1970, Mitchell et al (U.S.A.) 1971 and Rose (Canada) 1971.

At birth, diagnosis of congenital heart disease is difficult. Symptomatology of various congenital heart disease appears clearly at later age, moreover, benign murmurs are commonly heard at birth.

In order to know the incidence of various types of congenital heart diseases we classified our patients according to the classification given by Beverly, C. Morgan (1973). Acyanotic patients constituted the bulk i.e. 80.6% and the cyanotics were only 19.4%.

V.S.D. had been the most common type of congenital heart disease so far been observed by various workers. We also found 48.2% of our patients having V.S.D. while Keith et al (1974) reported it to be 28.3% in hospital of sick children, Toronto from 1950 to 1973, Krovets et al (Gainesville) 26%, Gassel (Chicago) 18%, NERICF 16.6% and Srivastava and Tandon (AIIMS, Delhi) 27%. The higher values in our set up can be because it is an easily diagnosable condition, and it is likely to be easily picked up for referral by practitioners of poorly equipped peripheral areas of Jhansi because of its loud murmur and thrill.
Roger in 1979 described few asymptomatic patients with cardiac findings similar to that of VSD. Since then all asymptomatic small ventricular septal defects have been called Roger’s disease. We noted 26.7% of our patients having small VSD, a finding comparable to 24.5% of Nadas and Fyler’s study. Keith reported it in 40% of his VSD cases. There is likely to be some dilution in our data as patients of this category are usually asymptomatic or have mild symptoms, they are usually not taken to hospital in our set up contrary to the routine regular check ups of children in developed countries.

Fifty percent of our patients of VSD fall in group having clinically large type of VSD. Its incidence reported by other workers were 82% (Wood) 36% (Bloomfield) and 50% (Nadas and Fyler).

The second common anomaly in our study was ASD comprising 12.9% of all patients. This is very near to various previous studies - 10.3% (Keith et al, 1974), 8% (Gassel) 6% (Krovetz et al) and 13% (Srivastava and Tendon) NERICP study however reported comparatively lower incidence i.e. 2.9%. But this study was conducted in infants and at this age ASD patients usually remain asymptomatic and many may remain undetected unless revealed in routine check up, which is a rarity in our set up.
Next in the occurrence was PDA, as 8.1% of our patients exhibited this type of defect. The figures from other workers ranged from 6.1% to 11% (Keith et al., 9.8%, Krovets et al. 11%, Gassel 11%, Srivastava et al. 11% and NERICP 6.1%).

Five (8.1%) patients from present study had tetralogy of Fallot. Keith et al. observed 9.7%, Krovets et al. 11%, NERICP 8.9%, Gassel 11%, Paulwood 11% and Srivastava et al. 17% of their patients. Our values are slightly lower than others but it will not be wise to compare them due to small sample size of our study.

Next in occurrence was transposition of great arteries, as 4.9% of our patients had it. Previous studies reported it to range between 4% to 5% by various workers (Keith et al., 4.9%, Gassel 4%, Krovets et al. 5%).

Male dominance in patients of congenital heart disease is a universally known fact. The male female ratio in our study was 1.9 : 1. Banerjee et al. (1978) also found it to be 1.7 : 1 in P.G.I., Chandigarh a figure very close to that recorded in the present study. Muir, Carlgren, MacMahon et al., Gardiner & Keith, Abbott, Roberts, Campbell and also in NERICP studies, male preponderance was noted. Thus it follow the general rule of more congenital anomalies of all types to be more common.
in males. Exception to this was PDA, where we found more girls than boys (40%). The female dominance in PDA was also noted by MacMahon et al (60%), Paulwood (70%), Keith et al (69%) and Campbell (73%).

VSD, Tetralogy of Fallot and transposition of vessels had higher male incidence of 70%, 100% and 67% respectively in our study. The male incidence for VSD was recorded to be 59% by MacMahon et al; for Tetralogy of Fallot were 61% (MacMahon et al) 60% (Keith et al) 64% (Paulwood) 59% (Campbell) and for TGA were 73% (MacMahon et al) 67% (Paulwood) and 66% (Keith et al). It could be by chance that all five patients of tetralogy in our study were male.

In case of ASD, we had equal number of children of both sexes. MacMahon et al also found similar results, but Keith et al and Campbell noted relatively more members of females than males i.e. 60% and 66% respectively.

In other groups of heart diseases, we can not comment upon sex ratio as there were very few patients in each group.

The incidence of other associated congenital defects are much higher in children with congenital heart disease than in general population (MacMahon et al, 1952; Campbell, 1968). Various figures from earlier studies varied from
16% to 20% i.e. (28% from HERICP studies 1968-1977, 25% by Greenwood et al, 1975, 21% by MacMahon et al, 1974 and 18% by Lamy et al, 1957). Surprisingly, the incidence of associated anomalies with congenital heart disease was low in our study i.e. 6.4%. Some chances of missing various internal abnormalities might be there as no specific investigation for different systems were undertaken except for heart.

Family history of congenital heart disease was present in 3 cases (4.8%). Among them one had same lesion as that of the patient i.e. VSD. McKenna et al (1958) found it to be 1.8%, Campbell (1965) 1.7% and Fuhrman (1961) 2.7%.

Several previous studies which linked Oestrogen/Pregesterone to congenital heart defects reported approximately twice the expected prevalence of heart defects among infants with prenatal exposure to exogenous female sex hormones in early pregnancy (Harlap et al, 1977, Hora et al 1976, Heinonen et al, 1977). Harlap et al, 1977 observed the risk of major malformations as major CNS, GIT or genitourinary malformation, cleft lip/palate, polydactyly, syndactyly, hip dislocation, heart diseases, Down syndrome, Glucose-6-phosphate dehydrogenase deficiency to be 20% higher in group exposed to nonexposed ones.
whereas for minor malformations as hypospadias, Inguinal/ or umbilical hernia, hydrocele, haemangiomas and Telangiactasia, the increase is about 33%. Part of this increase in risk may be due to tetratogenic effects of these hormones.

Janerich et al (1977) reported strong association with prevalence ratio of 6.5. Some positive association was also noted by Rothman et al., 1979. In our study, in 6.5% of patients, a positive history of Oestrogen/Progesterone intake was present.

History of taking antitubercular drugs in 2 of our cases can be a coincidental finding as no positive association to congenital heart defects is available in literature except for phenobarbitone and phenothiazine (Rothman et al., 1979, Heinonen et al., 1977).

In a single patient, history of overexposure to radiation during fourth month of gestation was available. This association can be either by chance or may have some importance as Cox (1964) noticed that all the malformations in general were twice as common in children of mothers, who had been exposed to frequent X-ray examination during gestation.

The most common presenting symptoms of cyanotic heart diseases were cough and breathlessness (46%), recurrent chest infection (36%), feeding difficulty (30%), failure to thrive (24%) and exertional dyspnoea (12%).
Chances of recurrent chest infection are more due to increased pulmonary circulation because of left to right shunts. Apart from this cough can also occur due to congestion of abdominal viscera from right heart failure, which can also cause dyspeptic symptoms as diarrhoea and vomiting (Friedman, 1984). Failure to thrive is a consequence of decreased systemic output, congestive heart failure leading to feeding difficulty and due to negative balance caused by recurrent chest infections.

Cyanotic heart disease patients presented with exertional dyspnoea (67%), cough and breathlessness (50%), cyanotic spells (41.6%), squatting (23%), feeding difficulty (29%) and failure to thrive (41.6%) in this study. Exertional dyspnoea occurs due to arterial unsaturation especially following exercise which increase oxygen demand. But this actually decreases on exertion because of increased right to left shunt, thus chemoreceptors get stimulated causing dyspnoea. An equally common cause of dyspnoea is due to increased pulmonary blood volume and increased pulmonary capillary pressure inherent in left sided failure. Cyanotic spells are said to be expression of cerebral anoxia resulting from drop in arterial saturation due to sudden spasm or right ventricular infundibulum resulting in precipitous drop in the pulmonary flow which usually follow a severe exertion. Franck (1963) described an other possible mechanism
presupposing that any stimulus that decreases blood and
tissue $\text{PO}_2$ and pH and/or raises the $\text{PCO}_2$ causes hyperpnoea,
which in turn increases systemic venous return. Since
pulmonary blood flow is fixed or decreased, an increased
volume of poorly oxygenated blood is shunted into aorta.
Gunteroth (1965) suggested that hyperventilation with
its consequent increase in venous return to the obstructed
right ventricle may be one of the underlying cause of
hypoxic spells.

However, the severity of these symptoms depends
upon degree of the pulmonic stenosis. Squatting, described
by Tausig is a characteristic posture assumed after
exertion or motionless standing with certain types of
congenital heart diseases especially tetralogy of Fallot.
This causes exclusion of highly unsaturated lower extremity
blood from circulation and augment the peripheral
resistance, thus diminish the degree of right to left shunt.
Vijay Priya et al (1979) noted presence of cyanotic spells
and squatting in one third of their tetralogy patients. In
our study 80% patients had cyanotic spells and 60% had
history of squatting.

Assessment of Development Quotient (D.Q.) is one of
the methods of expression of development, for the purpose
of comparison (Prabhakar et al). Thus on calculating develop-
mental quotient in each case, we found that there was significant retardation in motor field compared to the social and speech. The delay was more marked in the cyanotic patients. Possible explanations for the growth interference include malnutrition, tissue anoxia, diminished peripheral blood flow, chronic cardiac decompensation, genetic and endocrine factors and frequent upper and lower respiratory infections (Friedman, 1984).

Ruth et al (1982) studied developmental delay both in motor and mental development with congenital heart disease by means of Bayley scales of Infant Development and clinical neurological examinations. The abnormalities can be attributed to decreased arterial oxygen saturation, physical incompacity and psychological and emotional factors.

Feeding difficulties, negative balance and loss of appetite is due to recurrent chest infections and congestive heart failure. Decreased systemic output responsible for underdeveloped muscle mass and decreased physical activity because of that, are the important factors responsible for malnutrition. In cyanotics, unsaturated blood is an additional factor. One of the undernourished child in this study was also having caesarean, done for the high type of imperforate anus. In that case associated malabsorption was an exeptional factor.
As hemodynamic load is small in patients of small and moderate size VSD, these children tend to have a normal ECG. The shunting of blood occurs during systole at a time when right ventricle is also contracting and its volume is decreasing. Therefore, the shunted blood streams to pulmonary artery more or less directly, without any strain to right ventricle. On the contrary, this increased amount of blood passes through lungs and reaches left atrium and then in left ventricle. So when ventricular defects are without pulmonary arterial hypertension the ECG may show left ventricular hypertrophy. Later when either pulmonary stenosis or pulmonary arterial hypertension is developed, they show RVH and LVH or pure RVH.

Welch & Kinney (1942) pointed the association between left to right shunt and pulmonary vascular disease. By later studies also it has been established that by transmission of high pressure from left ventricle to the right ventricle or from aorta to pulmonary artery pulmonary arterial hypertension occurs. In these infants, there is persistence of fetal pattern of lung fields and involution of fetal arteriolar changes progress at a slower pace. However, individual susceptibility of arterioles and left atrial hypertension as in VSD also play a role in causation of pulmonary vascular disease. Commonly, it originates from combination of increased flow and increased resistance either singly or both.
Among the 4 cases of pure ASD, normal ECG was found in one and 3 out of 4 had RVH to accommodate and pump large amount of blood. rSR' pattern of right bundle branch block was present in 3 cases in right precordial leads representing delayed posterobasal activation of the ventricular septum. The patient having ASD with pulmonary stenosis had RVH.

In 3 cases both ASD and VSD were present, one of them was having evidence of RVH and remaining 2 of RVH. All these patients presented the picture of large VSD but after echocardiography only, associated ASD was disclosed to us.

Left axis deviation without any evidence of LVH was found in the single case of endocardial cushion defect having common atrio-ventricular canal.

All the cases of tetralogy of Fallot were having RVH. RVH was also found in cases of Ebstein anomaly, hypoplastic left heart syndrome and 2 out of 3 cases of TGA. Evidence of LVH was present in ECG in a case of TGA, a rare finding. Fortunately echocardiography was available with this patient showing pulmonary stenosis significantly obstructing the outlet of left ventricle. There was a single case of tricuspid atresia with LVH on ECG.
Radiological findings of pulmonary plethora were present in most of the cases of large VSD, ASD and PDA due to increased blood flow from left to right shunts. Plethora is a characteristic appearance of the lung vessels found in these condition and rarely from increase in cardiac output in certain other conditions. The main pulmonary artery is enlarged producing a convex pulmonary bay. Among 33% of cases of cyanotic heart disease, prominent pulmonary conus was there, but some cases, it could be concealed by big thymus in children. The pulmonary arteries and veins are increased in size and can be followed into the outer third of the lung. These findings of pulmonary arterial hypertension has to be differentiated from various causes of pulmonary venous hypertension, produced by back pressure to pulmonary veins because of impairment of functions of valves or chambers. In that case distension of normally collapsed upper lobe veins are seen. Later early A and B lines, interstitial oedema, alveolar pulmonary oedema (Sat's wing appearance) and pleural effusion may develop (By Raphael & Donaldson in Textbook of Radiology). Characteristic shape of heart i.e. Boot shaped heart was seen in 3 out of 5 cases, of Fallot's tetralogy in this study. This is due to RVH which lifts up relatively hypoplastic left ventricle and because hypoplastic pulmonary artery. About 33% of patients of Fallot's also have right aortic arch (Pearson & Rigby in Textbook of Radiology). We
found it in 20% of cases. Egg shaped cardiac shadow was present in 1 out of 3 cases of TGA which occur because of absence of pulmonary artery at its conventional site, RVH and narrow superior mediastinum. However, in other cases of TGA and Fallots, right ventricular type of cardiomegaly was found.

Out of 50 of the acyanotic group, in 31 patients where echocardiography was possible, diagnosis made out after clinical examination, ECG and X-ray chest remained the same in echo in all of the cases. But then some additional cardiac anomalies were detected, as in 4 cases where we suspected only VSD clinically, both ASD and VSD were seen in 3 cases and in the rest one case, PDA was also present with it.

One case needs elaboration here, which was getting cyanosed while crying and was having signs of ASD. We suspected him to have some cyanotic heart disease, but after echo ASD was found to be present alone without any associated anomaly. The cause of cyanosis was then thought to be right to left shunt occuring during cry. Because of this reason congenital heart diseases are classified in two groups, one having cyanosis and other with little or no cyanosis, as mentioned in Nelson Text book of Pediatrics.
Another case, which was diagnosed as of ASD by clinical examination, ECG, X-ray chest and also by echocardiography, marked clubbing was found on examination and cyanosis was not present at any time. The child was also having imperforate anus for which colostomy was made. So it was interpreted that clubbing in that case was of non-cardiac origin probably due to chronic malabsorption because of long standing (2 years) colostomy.

In cases where small VSD was suspected, echo revealed small VSD in 6 out of 8 cases. Doppler studies are needed for such cases as very small defects can be missed by 2D & M mode echocardiography.

In cyanotic children except for tetralogy of Fallot which could be diagnosed fairly well by clinical examination, ECG and X-ray chest for all other cases echocardiographic studies were found to be useful in diagnosis. For example, a four month old child presented to us with feeding difficulty and cyanosis since birth. ECG showed left atrial overload only and X-ray chest showed Cardiomegaly (left ventricular type). We were no where near the diagnosis. Later, in follow up patient showed the Echo report confirming left atrial and left ventricular dilation. Contrast dye when injected to systemic vein reached in left atrium instead of right and from there to left ventricle and right atrium via ASD. So
a diagnosis of anomalous systemic venous drainage was made, which is a rare condition. In the same patient family history suggestive of some cyanotic heart disease was there in 2 of his siblings.