# A Study of Congenital Heart Disease in Karachi, Pakistan

*By*

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Reports on congenital heart malformation in populations of Indo-Pakistan origin are scanty and analysis of congenital cardiovascular malformations in the South Africans Indians failed to reveal any difference from other reported series of caucasian countries in the incidence and type of congenital heart disease. This report is based on our experience with congenital heart disease and is undertaking to provide reliable information on this subject in this racial group.

## Material and Methods

The observations on which this study is based were made on 230 patients with suspected congenital heart disease, who attended the congenital heart clinic of this Centre. The series was closed at the 230th case, for by then 200 cases of proved congenital heart disease were collected.

Table I shows the breakdown of the 230 cases. Fifteen had functional murumurs, two were found to have rheumatic heart disease, in both cases mitral incompetence. The remaining 14 patients failed to complete investigations which were necessary for the elucidation of the nature of their signs and symptoms and therefore a definite diagnosis could not be made. Taking into consideration the very poor follow up rate in the hospital patients in our country, this incidence of 7% defection is infact very low.

The ages of the patients ranged between one month and 65 years with a mean age of 9.2 years. The male to female ratio was 3:2. This sries is unique in the respect that (the study has been done on all age groups. Wood's series and that of Somerville et al., did not include infants upto one year and had fewer number of children. The series of Keith, Rowe and Vlad and of Nadas included patients below the age of 14 years only. The majority of subjects in the study of Van der Horst et al., were adolescent and young adults between 10 and 25 years of age.

The diagnosis was confirmed by cardiac catheterisation in 37 cases, by additional help of angiocardiography in six cases and during surgery in seven cases. No autopsy could be performed due to non-cooperation of the relations of patients. Nevrtheless we do hope to present some autopsy series in coming years as the ice in this regard has just started breaking.

The procedures of cardiac catheterisation and angiocardiography were mainly of diagnostic value in our series and did not contribute to the treatment of the patients. Since these procedures have their inherent morbidity and mortality the decisions to catheterise induvidual cases were based upon three considerations: (a) safety of the procedure: patients who were either too till or

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too young were not catheterised; (b) nature of the lesion: straight forward cases where a definite diagnosis could be made by clinical means alone were not catheterised, whereas cases with complicated and multiple lesions were catheterised more often; and (c) availability of surgical repair: cases suspected to be suffering from a defect amenable to the surgical facilities available, were catheterised more often than cases for whom surgical help is not yet available in our Centre. This policy was also adopted by Van der Horst et al. We must stress that the last fact only influenced our decision to perform cardiac catheterisation but did not have any effect on overall patient population of this series and we are reasonably certain that this series did not include a greater number of cases amenable to surgical repair.

## Results

Figure I gives the relative incidence of different congenital amomalies found in our series. Ventricular septal defect was the commonest lesion in this series. The five major congenital heart defects the ventricular septal defect, the atrial septal defect, the patent ductus arteriosus, the pulmonary stenosis and the tetralogy of Fallot comprised more than 80% of the entire series.

*Ventricular septal defect:*

Table II shows the details of the cases with ventricular septal defect. There were 42 cases comprising 21% of the entire series. Male to female ratio was 9:5. Ages ranged between one month and 25 years with a mean age of 5.6 years. The majority of cases were between the ages of 0 to 10 years. There was no case above the age of 30 years.

*Atrial septal defect:*

Table III gives the details of the cases with atrial septal defect. There were a total of 32 cases comprising 16% of the entire series. Male to female ratio was surprising 9:7. Ages ranged between one month to 65 years with (a mean age of 16.5 years. The majority of patients belonged to 0 to 10 age group. There were 3.1% cases above the age of 60 years.

*Patent ductus arterious:*

Table IV gives the details of the patients with this defect. There were a total of 24 cases forming a percentage of 12. Male to female ratio was 1:2. Ages ranged between 3 months and 29 years with a mean age of 6.5 years. The majority of the patients again belonged to 0 to 10 age group and there was no case above the age of 30 years.

*Fallot's tetralogy:*

Table V gives the details of the cases with this defect. There were a total of 33 cases forming 16.5% of the entire series. Male to female ratio was 7:4. Ages ranged between 3 months and 23 years with a mean age of 8.2 years. Again the majority of cases belonged to 0 to 10 years and there was no case above the age of 30 years.

*Pulmonary stenosis:*

Table VI gives the details of the cases with this defect. There were a total of 31 cases forming 15.5% of the entire eseries. Male to female ratio was 1.6:1. Ages ranged between 2-1/2 months and 47 years with a mean age of 9.6 years. Again the amjority of cases belonged to 0 to 10 years. There were 6.4% cases above the age of 30 years.

Other defects:

Relatively rare congential defects like transposition of the great vessels, tricuspid atresia, atrial septal defect with pulmonary stenonsis, persistant truncus arterious and Eisenmenger's Syndrome etc., comprised 8% of the entire series. Surprisingly there were only two cases (1%) of coarctation of aorta.

Table VII gives a summary of haemodynamic data of the 47 patients studied.

## Discussion

Three important series of congenital heart disease have been reported on caucasian races. Wood reported his series of 900 cases. This was enlarged to 2000 cases by Somerville et al. Keith, Rowe and Vlad reported 6647 cases. Nadas's series comprised of 3786 cases. Report of Van der Horst et al., is the only reliable one on population of Indo-Pakistan racial origin. Table VIII shows the relative Frequency of different congenital heart defects as found in our series and those reported by other workers. Our series closely resembled the one reported by Nadas where the ventricular septal defect was the commonest lesion. In this report our series also resembled that of Keith, Rows and Vlad, who reported 25% incidence of ventricular septal defect, but it differs very significantly from that of Wood and Somerville et al., who reported 8% and 12% incidence of ventricular septal defect respectively. The commonest lesion in Wood's series was atrial septal defect. The tetralogy of Fallot was the commonest lesion in Wood's first 200 cases of congenital heart disease. The second commonest lesion in our series was tetraolgy of Fallot as was also in the series of Wood and that of Nadas. The second commonest lesion in the series of Keith, Rowe and Vlad was patent ductus arteriosus. Our series also closely resemble the series of Van der Horst et al., except in the incidence of tetralogy of Fallot, pulonary stenosis and Eisenmenger's Syndrome which were encountered most frequently and that oviously which were encountered less frequently in our series. This incidence of isolated coarctation of aorta was identicle. This is considerably low as compared to Western figures.

The differences in the relative frequencies of different congenital lesions between our series and the series reported by other authors are, in our opinion, the result of the difference in the ages of the patients studied. Wood's series and that of Somerville et al., did not include infants and they mainly comprised of adults. On the contrary the series of Keith, Rowe and Vlad and Nadas did not include patients above the age of 14 years and were entirely based on children material. Our series included patients of all age groups.

It appears that younger the average age of the patients of a particular series, the higher the incidence of ventricular septal defect (Keith, Rowe and Vlad series) and older the average age of the series, the higher the incidence of atrial septal defect (Wood series). Our series though comprising of all age groups of patients, still have ventricular septal defect as the commonest lesion, but the incidence of atial septal defect is also comparatively high at 16% as compared to that of Nadas of 10.04% and of Keith, Rowe and Vlad of 7%. The higher incidence of atrial septal defect in older patients population is a reflection on the relatively benign nature of the defect.

There are significant differences in the incidence of Eisenmenger's Syndrome in different series.

We had only two cases (1%) the reversal of the shunt being at the patent ductus artrious level in one and at the ventricular septal defect level in the other. Wood reported an incidence of 6.9% in his 900 cases whereas the incidence of this syndrome reported by Nadas was more than 10%. Van der Horst reported an incidence of 2.9%. To our mind there could be 3 possible explanations for the low incidence of Eisenmenger's syndrome in our series.

(1) *Persistence of foetal type of pulmonary vasculature:*

An increased pulmonary blood flow can prevent the regression of the foetal type of pulmonary arterioles, resulting in early right ventricular failure and death. This would result in a mortality in early infancy, so that only a few cases would live long enough to be seen by the workers. An extra-ordinarily severe reaction of pulmonary vasculature to raised left atrial presure in mitral stenosis has been shown in this country by Abbasi et al.

(2) *Hyporeactive pulmonary vasculature:*

The pulmonary vasculature may be hyporeactive to increased flow with the result that a very small proportion of patients with left to right shunt develop pulmonary hypertension. Hence only a small number of patients would reverse the shunt. This possibility appears rather unlikely.

(3) *The effect of the size of the series:*

Table IX shows the relative incidence of Eisenmenger's syndrome in different series. Wood's first 200 cases had an incidence only 1% which is the same as in our series. The incidence of this syndrome rose to 6.5% in Wood's 900 cases and it further rose to 8% when he reported his 1000 cases. In a series of 3786 cases of Nadas the incidence of the syndrome was 10%. It appears that when a group of workers start their study of congenital heart disease they attract all cases without any bias to a particular type of defect, but when they acquire a higher professional status, a large proportion of relatively complicated and surgically incurable cases like Eisenmenger group get referred to them, with the result that the relative incidence of these type of cases rises in their series.

Except for minor differences the spectrim of cardiac malformation reported in our study is similar to other reports on population of caucasian and Indo-Pakistan racial origin. The minor differences are mainly the result of the differences in the age groups of patients in different series. The experience, special interest of the workers and the frequency with which sophisticated diagnostic tools were used also appear to influence the reported incidence of different congenital heart disease malformations. Racial origin does not seem to influence the incidence of congenital heart defects.

## Summary

A total of 230 patients with suspected congenital disease were studied, out of which 200 cases of proved congenital heart disease were found. The series comprised of all age groups of patients from one month to 65 years with a mean age of 9.2 years. Ventricular septal defect was found to be the commonest congenital heart lesion, followed by tetralogy of Fallot and atrial septal defect. A comparison is made with other reported series of Wood, Somerville et al., Keith, Rowe and Vlad, Nadas and Van der Horst et al. The incidence of different cardiac malformation does not differ from other reported series on population of caucasians and Indo-Pakistan racial origin.

Table I Details of the Cases Studied

| | |
|---|---|
| Total No. of Cases Studied | 230 |
| Functional Murmurs .. .. | 14 |
| Failed to Attend for follow up investigations .. .. .. | 13 |
| Rheumatic .. .. .. | 2 |
| Cardiomyopathy .. .... | 1 |
| Proven cases of congenital heart Disease .. .. .. | 200 |
| Total: | 230 |
| Diagnosis confirmed by catheterization .. .. .. | 47 |
| Diagnosis confirmed by Angiocardiography .. .. .. | 6 |
| Diagnosis confirmed on Operation | 7 |
| Age Range 1 Month—65 Years | |
| Mean Age 9.2 Years | |
| M:F 3:2 | |

Effect of the size of series on the incidence of Eisenmenger syndrome

| *Author* | *Total No. cases of congenital Heart Disease* | *Eisenmenger syndrome* 1 |
|---|---|---|
| Wood (1950) | 200 | 1 |
| Wood (1956) | 900 | 6.5 |
| Wood (1958) | 1000 | 8 |
| Nadas (1963) | 3786 | 10 |
| Van der Horst et al. (1970) | 395 | 2.9 |

*Table VII:* Comparative Incidence of Major Congenital Heart Lesions in Literature and Our Series

| | *This* 200 | *Nadas* 3786 | *Keith* 6647 | *Paul Wood* 2000 |
|---|---|---|---|---|
| VSD | 21 | 19.97 | 25 | 12% |
| Fallots | 16.5 | 14.5 | 10.2 | 9.5 |
| ASD | 16 | 10.04 | 10.6 | 23.5 |
| PS | 15.5 | 11.97 | 8.5 | 23 |
| PDA | 12 | 12.31 | 12.1 | 9 |
| AS | 6 | 5.73 | 5.5 | 6 |
| VSD with PS | 3.5 | *N.M. | *N.M. | 1.5 (1954) |
| Transposition | 1.5 | 3.96 | 5.4 | 1 |
| Tricuspid atresia | 1.5 | 1.24 | 2.0 | .8 |
| ASD with PS | 1.5 | *N.M. | *N.M. | 2.5 |
| Coarctation | 1 | 4.99 | 5.6 | 10 |
| Truncus | 1 | .5 | .41 | Rare |
| Eisenmenger | 1 | 10 | *N.M. | 7 |

*N.M.: Note mentioned
* Reported by Somerville et al. 1968.

*Table II*. Ventricular Septal Defect

No. of Cases 42 Percentage of Total Cases 21 M.F. 9:5
Age Range 1 Month 25 Years Average Age 5.6 Years.

| Age group .. .. | 0—10 | 11—20 | 21—30 | 31—40 | 41—50 | 51—60 | Over 60 |
|---|---|---|---|---|---|---|---|
| VSD% .. .. | 83.3 | 9.5 | 7.1 | | | | |

*Table III:* Atrial Septal Defect

No. of Cases 32 Percentage of Total Cases 16 M.F. 9.7
Age Range 1 Month-65 Years Average Age 15.5 Years

| Age group .. .. | 0—10 | 11—20 | 21—30 | 31—40 | 41—50 | 51—60 | Over 60 |
|---|---|---|---|---|---|---|---|
| ASD% .. .. | 46.8 | 31.2 | 9.3 | 3.1 | 6.2 | 0 | 3.1 |

*Table IV*. Patent Ductus Arteriosus

No. of Cases: 24 Percentage of Total Cases: 12 M.F. 1:2
Age range 3 Month—29 Years: Average Age: 6.5 Years

| Age Group .. .. | 0—10 | 11—20 | 21—30 | 31—40 | 51—60 | 51—60 | Over 60 |
|---|---|---|---|---|---|---|---|
| PDA .. .. | 79.1 | 16.6 | 4.1 | 0 | 0 | 0 | 0 |

*Table V:* Tetralogy of Fallot

No. of Cases 33 Percentage of Total Cases 16.5 M.F. 7.4
Age Range 3 Months—30 Years Average Age 8.2 Years.

| Age Group .. .. | 0—10 | 11—20 | 21—30 | 31—40 | 41—50 | 51—60 | Over 60 |
|---|---|---|---|---|---|---|---|
| Fallots% .. .. | 66.6 | 27.2 | 6.00 | 0 | 0 | 0 | 0 |

*Table VI*. Pulmonary Stenosis

No. of Cases: 31 Percentage of Total Cases 15.5 M.F. 19:12
Age Range 21/2 Month—37 Years Average Age: 9.6 Years

| Age Group .. .. | 0—10 | 11—20 | 21—30 | 31—40 | 41—50 | 51—60 | Over 60 |
|---|---|---|---|---|---|---|---|
| PS% . | 70.9 | 16.1 | 6.4 | 3.2 | 3.2 | 0 | 0 |

Table VIII - Comparative Incidence of Major Congenital Heart Lesions in Literature and Our Series

| Authors and Total Patients | This Series 200 | Nadas 3786 | Keith*** 6647 | Paul Wood** 2000 | Van der Horst et. al. |
|---|---|---|---|---|---|
| Ventricular Septal Defect | 21 | 19.97 | 25 | 12% | 26.3 |
| Fallots | 16.5 | 14.5 | 10.2 | 9.5 | 11.4 |
| Atrial Septal Defect | 16 | 10.04 | 10.6 | 23.5 | 17.2 |
| Pulmonary Stenosis | 15.5 | 11.97 | 8.5 | 13 | 7.1 |
| Patent Ductus Arteriosus | 12 | 12.31 | 12.1 | 9 | 14.4 |
| Aorta Stenosis | 6 | 5.73 | 5.5 | 6 | 4.7 |
| Ventricular Septal Defect with Pulmonary Stenosis | 3.5 | *N.M. | *N.M. | 1.3 | — |
| Transposition | 1.5 | 3.96 | 5.4 | 1 | 3 |
| Tricupid Atresia | 1.5 | 1.24 | 2.0 | 0.8 | 1.4 |
| Atrial Septal Defect with Pulmonary Stenosis | 1.5 | *N.M. | *N.M. | 2.5 | — |
| Coarctation | 1 | 4.99 | 5.6 | 10 | 1% |
| Truncus | 1 | 0.55 | 0.41 | Rare | — |
| Eisenmenger | 1 | 10 | *N.M. | 7 | 2.9% |

*N.M.: Not mentioned. **Reported by Somerville et al. 1968. ***Keith et al.

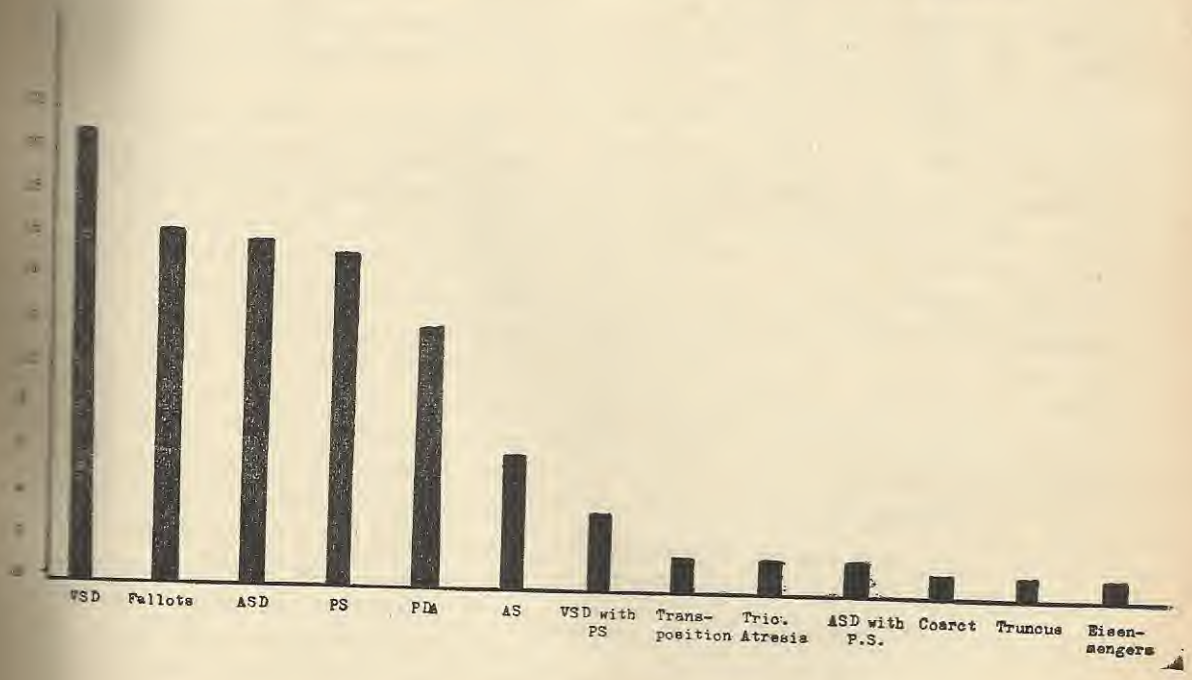


Fig.1.
Relative Incidence of different Congenital Anamolies.