

The Prevalence Of Congenital Heart Diseases Among Down's Syndrome Children In Diwaniyah Governorate

**Mohanad Al-Jashami *AL-Diwiniyah medical office*

ABSTRACT :

Objective :A survey was carried out to assess the prevalence of congenital heart diseases (CHD).

Patients And Methods: among (180) Down's syndrome (DS)children in institute for mentally handicapped (Alraga institute) and outpatients referred for ECHO study in Diwaniyah Teaching Hospital.

RESULTS: It was found that (61.1%) of DS cases studied had CHD with a M:F ratio of 1.5:1. The commonest type of CHD found was ventricular septal defect (VSD), followed by atrial septal defect (ASD), mitral valve prolapse(MVP), tetralogy of fallot (TOF), patent ductus arteriosus (PDA) and co-arctation of aorta in (54.5 %, 17.2 %, 14.5 %, 11.8 %, 0.9%, 0.9% respectively). The most common mode of presentation of CHD was recurrent chest infection. It was found that about 17.5% of cases with VSD had spontaneous closure, while only (7.5%) were treated surgically. The rest are left untreated.

INTRODUCTION:

The incidence of DS is around 1in 700 of all live births, however it varies with mothers' age where it is 1 in 1400 among infants born to 25 years old mothers, and 1 in 46 among infants to 45 years old mothers. CHD occur in (40-60)% of infants with DS⁽¹⁾. VSD and ASD accounting for approximately (71.3%) of lesions.

Cardiac defects are of special significance in DS, since they are the main factors that determine life expectancy Large left to right shunts in DS seem to progress to pulmonary hypertension more quickly than in normal children and sever cyanosis and shunt reversal may be seen very early⁽³⁾.Early diagnosis and corrective surgery may improve survival rates to (80-90)% of children who may otherwise fail to reach their 15th year⁽⁴⁾. This study aims at

determining the prevalence and type of congenital heart disease among children with Down's syndrome in institutes for the mentally handicapped and referred out patients for ECHO examination in Diwanayah teaching hospital.

PATIENTS AND METHODS:

Across-sectional study was carried out in institute for the mentally handicapped (Alraga instate),and outpatients referred for ECHO examination in Diwanayah teaching hospital from August 2003 to May 2006. All cases affected by DS and they were between (3months- 18 years) of age. History, general information, clinical examination as well as investigations results were all recorded in specifically arranged data collection forms. The investigations used to diagnose CHD included ECG, CXR and ECHO.

RESULTS AND DISCUSSION:

Table 1 shows that about (61.1%) of DS cases have congenital heart diseases, while other studies show these prevalence such as 40% in Rowe Uchida study (1961) in Canada⁽⁵⁾. 62.3% in Greenwood and Nada's study (1976) in Boston, USA⁽⁶⁾; 48% in Wells et al study(1994) in Birmingham, UK⁽⁷⁾.; 38.7% in Garcia et al study (1997) in Spain⁽⁸⁾; 44% in Freeman, et al study (1998) in Atlanta, USA⁽⁹⁾; 56% in Paladin et al study (2000) in Italy⁽¹⁰⁾; 41% in Al-Arrayed and Rajab study (1995) in Bahrain⁽¹¹⁾. 36% in Hussein study (1999) in Mousl, Iraq⁽¹²⁾, and 53.3% in Abd-Al-Mohsin study (2000) in Baghdad, Iraq⁽¹³⁾. The different prevalence's in those studies could be due to differences in subjects selection based on hospital data or those samples with ages below 4 years.

In the present study, the relatively higher prevalence of CHD is due to

the selection of the sample from specialized instute for mentally handicapped and outpatients referred for ECHO examination.

Table 2 shows that of those 110 cases affected by CHD ,the majority were males with M:F ratio of 1.5:1, this is not consistent

with results obtained by Breg et al (1960) from London showing M:F ratio of 0.9:1⁽¹⁴⁾; and by Tandon and Edwards (1973) from USA with a M:F ratio of 0.83:1⁽¹⁵⁾. This variation could be explained on the basis of different sampling techniques or social factors where males are being better cared for in our community.

Table 3 shows that the majority (54.5%) of DS cases with CHD have VSD similar to results obtained for non-DS,CHD children in Baghdad⁽¹⁶⁾, followed by ASD in 17.2% of cases; MVP and TOF were found in 14.5% and 11.8% of cases respectively. The least common types of CHD were PDA and Co-arctation of aorta which occurred in only one case each (0.9%), a result similar also to that obtained for non-DS,CHD children in Baghdad⁽¹⁶⁾.

The rates of different types of CHD in DS cases varied among different studies, this variation in rates is usually small except for Warkany et al study (1966) where markedly higher rates were seen for VSD,ASD , MVP and TOF⁽¹⁷⁾.

The variation could be explained on the basis of different criteria of the study groups. In the present study ,VSDs are more predominant than ASDs and this is consistence with other studies^(11, 12,17,18,19,20), while the opposite was found by others^(5,6,7,9,13,15).

Table 4 shows that 55% of cases with CHD had developed signs and symptoms of CHD within the first months of life, and that 30% of cases of DS with CHD had given no physical signs or symptoms and were only diagnosed accidentally. This is consistence with results obtained by Laursen (1976) in Denmark⁽¹⁹⁾. No other studies regarding this feature could be found. The early onset of symptoms and signs in DS children with CHD is because they are more liable to recurrent infections and early development of cardiac and pulmonary complication than do non-DS children with similar cardiac anomalies⁽¹⁹⁾.

Table 5 shows that the most frequent clinical manifestations of CHD in DS cases was recurrent chest infection similar to results obtained for a group of non-DS,CHD children in Baghdad, but to a much higher rate⁽¹⁶⁾, followed by cough, dyspnea and fever in

82.5% ,80%, 70%, and 60% respectively .The least common were clubbing and heart failure, this finding is consistent with the results obtained by Shaher et al (1972) in Albany⁽²¹⁾. Such results support the fact that infections of the respiratory tract are common complications in patients with DS, both with and without CHD⁽¹⁹⁾. These respiratory problems are related to many structural and functional disorders associated with DS ,and as a result of mild immune deficiency⁽⁴⁾.

Table 6 shows that 17.5% of cases had VSD that closed spontaneously and 7.5% were surgically dealt with , the majority(75%) were left untreated. Tandon and Edwards (1973) in USA⁽¹⁵⁾ show that only 1.8% of VSD in their study group has spontaneous closure which is much lower than the rate obtained in this study. This variation could be due to differences in the age structure of the two study groups .Other available studies showed no mention of spontaneous closure.

The majority of children (75%) in this study were untreated which is a high percentage probably due to lack of interest of many surgeons in operating upon such children and also lack of interest of families of DS children to look after them, perhaps because of unsatisfactory consequences such as complications and unavailable nursing facilities, while Okada et al (1993)⁽²²⁾ in Japan have suggested that early diagnosis and surgical repair were the key elements in the management of patients with DS and VSD, a suggestion supported by Reller and Morris (1998) in Oregon, USA who noted that the survival outcomes after surgery for cardiac malformations were similar for children with and without DS⁽²⁴⁾.

CONCLUSION

In this study shows that CHD affects (61.1%) of DS children with predominance of VSD, with males being affected more than females .

Symptoms and signs of CHD appear in the early months of life which reflect the severity of CHD. Recurrent chest infections being the most common clinical manifestation. The majority

(75%) of DS children are left untreated due to lack of facilities or interest.

Table 1. Down Syndrome cases By CHD

DS cases	Number of cases	%
With CHD	110	61.1
Without CHD	70	38.8
Total	180	100.0

Table 2. Down Syndrome cases with CHD By Gender

Gender	Number	%	M:F
Males	66	60.0	1.5: 1
Females	44	40.0	
Total	110	100.0	

Table 3. Down Syndrome Children With CHD By Type Of CHD

Type of Defect	Number	%
VSD	60	54.5
ASD	19	17.2
MVP	16	14.5
TOF	13	11.8
PDA	1	0.9
Coarctation of Aorta	1	0.9
Total	110	100.0

Table 4. Onset Of Symptoms and Signs Of CHD Among Down's Syndrome Children

Onset of symptoms and signs	Number	%	
At birth	14	12.7	55%
1st weeks	16	14.6	
1st months	30	27.2	
1st year	6	5.4	
>1 year	12	10.9	
No signs & symptoms	34	30.0	
Total	110	100.0	

Table 5. Down Syndrome cases With CHD By Signs and Symptoms

Signs and symptoms	Number	%
Recurrent chest infection	33	82.5
Cough	32	80.0
Dyspnea	28	70.0
Fever	24	60.0
Cyanosis	22	55.0
Changing of feeding pattern	22	55.0
Failure to thrive	16	40.0
Reluctance to feed	9	22.5
Clubbing	6	15.0
Heart failure	4	10.0

Table 6. Down Syndrome cases with CHD and Result of their management

Results	Number	%
Closed spontaneously (VSD)	19	17.5
Correction by surgery	8	7.1
Untreated	83	75.4
Total	110	100.0

REFERENCES:

1. Patton M.A. Genetics. In; Forfar and Arneil's Textbook of Pediatrics. 5th Edition. AGM ,and McIntosh N (Editor). Churchill Livingstone, New York, 1998, pp.54-55.
2. Houston A.B. Cardiovascular Diseases. In: Fofar and Arneil's Textbook of Pediatrics. 5th Edition. Campbell AGM, and McIntosn N (Editors). Churchill Livingstone, New York, 1998, p596.
3. Hall D.M.B. and Jolly H. in: The Child with a Hadicap. 1st Edition. Blackwell Scientific Publications, Oxford , 1984, pp. 197-198.

4. Cody H. and Kamphaus R.W. Down syndrome. In :Handbook of Neurodevelopmental and Genetic Disorders in Children. 1st Edition Goldstein S. and Reynolds C. R. (Editors). The Guilford Press, New York, 1999,pp.387-390.
5. Rowe R. D. and Uchida I.A. Cardiac Malfunction in Mongolism : a prospective study of 184 Mongoloid children. AM. J .Med; 1961, vol.31, pp. 726-735.
6. Greenwood R.D. and Nadas A. S. The Clinical Course of Cardiac Disease in Down's syndrome.
7. Wells G.L., Barker SE., Finely S.C., Colvin E.V. and finely W.H. Congenital Heart Disease in infants with DS. South Medical JOURNAL, 1994, Vol.87, pp.724-727.
8. Garcia- Minaur S., Castro-Laiz v and Galdeano-Mirando J.M. Down's syndrome in the Baque Autonomous Community, 1990-1995: (Types of birth and follow-up of a cohort of 116 children during the first year of life. Registry of Anomalies) Esp. Peadiatrics 1997, Vol. 47, No. 1 pp.61-65 (Medline).
9. Freeman S.B., Taft L.f., Dooley K.J., Allran K., Sherman S.L Hassold T.J., Khoury M.J., and Saker D.M. Population-based study of congenital heart disease defect in DS. AM. J.Med. genet.,1998,vol . 80, 3 , pp.213-217(Medline).
10. Paladini D., Tartaglione A., Teodoro A., Forleo., Borghese A., and Martineli P. The association between congenital heart disease and down syndrome in prenatal life .Ultrasound-obstet. Gynecol., 2000, vol. no. 2 pp. 104-108(medline).
11. Al-Arrayad s. and Rajab K.E. Morbidity of down's syndrome among hospital population of patients in Bahrain . Journal of the Bahrain Medical society, 1995, vol. 7 no. 1, pp. 13-16. (extramed)
12. Hussien Y.A. A study of down syndrome in mousl. A Dissertation submittrd to the Iraqi Commission for medical specialization-pediatric, 1999.
13. Abd-Al-Mohsun E. study of congenital heart disease in down syndrome. A Disseratation submitted for degree of Diploma in child health,2000.
14. Berg J.M., Cromal., and France N.E. congenital cardiac malformations in mongolism. Br. Hr j. 1960, vol 22,pp. 331-346.

15. Tandon r., and Edwards J.E. Clinicopathologic correlations : cardiac malformation associated with down's syndrome. *Circulation*, 1973, vol. xl v11 pp. 1349-1355.
16. Al-Ward NJA and abd NS: Congenital heat disease in children 1: some.
17. Pidemiological and clinical findings. *J.Fac.med. Baghdad* (1999) vol. 40 no. 3.
18. Warkany J., Passarge E., and Smith L.B. Congenital malformations in autosomal trisomy syndromes. *AM. J. dis .child.*, 1966, vol. 112, pp.502-517.
19. Hassan i., Haleem A., and Bhutta Z.A. Down syndrome in Pakistan: presentation and prevalencs in a defined birth cohort. *Pakistan'sJ. Med sci.*, 1996, vol .12, no. 3, pp.207-211.(extramed).
20. Laursen H.B. Congenital heart disease in down's syndrome. *Br.hr. j.* 1976,vol.38,pp.32-38.
21. Cullum L., and Liebman J. The association of congenital heart disease with down;s syndrome (mongolism). *The American journal of cardiology* 1969,vol. 24 pp.354-357.
22. Shahar R.M., Farina M.A.,. Porter I.H., and Bishop M. Clinical aspects of congenital heart disease in mongolism. *The American J,of cardiology*, 1972, vol. 29 ,pp. 479- 503.
23. Okada H., Tsuboi H., Nishik., Matsumoto N., Gohra H., Katou., Fujimura Y., Miyamoto M.,and Esatok. Surgical treatment of VSD associated with down syndrome *kyobugeka*, 1993,vol. 46, no. 5, pp.369-398.(medline).
24. Kawai T., Wada Y., Enmoto T., Nishiyama k.,kitaura k., sato s. and oka T. Comparison of hemodynamic data before and after corrective surgey for down's syndrome and VSD. *Heart vessels*,1995, vol. 10, no. 3 pp.154-157 (medline).
25. Reller M.D., and Morris C.D. Is down syndrome a risk factor for poor outcome after repair of congenital heart disease .*J Pediat.*, 1998,no. 4,pp 738-741 (medline_.