

The spectrum of congenital heart defects in infants with down's syndrome, Khartoum, Sudan

Abstract

The aim of this study was to determine the prevalence and the types of congenital heart defects (CHD). In children with Down's syndrome (DS); and to assess the impact of CHD on nutritional status. This was a prospective cohort study of infants with DS over a six month period starting Sept 2011. CHD occurred in 43.1% of 109 children with DS. Atrioventricular Septal defect (AVSD) was found in 32%, Ventricular Septal defect (VSD) in 26%, Patent Ductus Arteriosus (PDA) in 17% and Atrial Septal Defect (ASD) in 15% of the affected infants. Over all under-nutrition was found in 62 (56.9 %) infants; 87.2% were in the CHD group and 33.9% in the group with normal hearts. The presence of CHD in children with DS had an influence on their nutritional status. Early recognition of the cardiac condition of neonates with DS by screening is justified.

Keywords: Sudan, down's syndrome, prevalence, congenital heart defects, infants, nutritional status

Volume 2 Issue 5 - 2015

Osama El-Shazali, Hamed Ahmed, Hafiz El-Shazali

Department of Pediatrics, University of Khartoum, Sudan

Correspondence: Osama El-Shazali, Department of Pediatrics, University of Khartoum, P.O. Box 102 Khartoum, Sudan, Tel 25000000000, Email osamaelshazali@hotmail.com

Received: July 21, 2014 | **Published:** August 31, 2015

Abbreviations: DS, down's syndrome; CHD, congenital heart defects; AVSD, atrioventricular septal defect

Introduction

Down's syndrome (DS) is the most common chromosomal malformation affecting about 1:700 live born. About half of children with Down's syndrome are born with congenital heart disease.^{1,2} CHD is considered to be the most important malformation of DS causing morbidity and mortality.³⁻⁵ In view of this, cardiac and echocardiography assessment of all newborns with Down's syndrome is the standard recommendation e.g., American Academy of Pediatrics.⁶ Previous studies by Freeman and colleagues had shown that ethnic differences in the types of CHD in Down's syndrome do exists, with a higher incidence of AVSD in blacks compared to whites and Hispanics.⁷ A study from Guatemala by Vida and colleagues has shown that the most common single defect was PDA, found in 54 of the 189 patients (28.6 %), and followed by VSD in 27.5 %, ASD in 12.7 % and AVSD in 9.5 %.⁸ DS is associated with impaired growth.⁹⁻¹² The growth of DS children with CHD is more significantly affected.^{13,14} In the present study we will describe the incidence and the types of CHD in DS children in Sudan as limited data exists about this important health issue. We will also look at the association of CHD with the nutritional status.

Method

This is a Cross-sectional descriptive study of the spectrum of congenital heart defects in infants with DS. All children seen in all the Khartoum state neonatal units and children hospitals with a clinical diagnosis of DS during the six months study period starting from September 2011 were included in the study. Only children less than 12 months of age were included. Infants known to have congenital heart disease were excluded from the study. Verbal informed consent was obtained from all the parents. A pre tested questionnaire was filled (annex) and all children underwent a detailed clinical examination and an echocardiography examination. All the echocardiography examination were done by one person using GE vivid 5 (GE Waukesha, WI). For the purpose of the study we only included aerodynamically significant lesion e.g. Patent Foramen Ovule (PFO), tiny closing PDA

was excluded. The statistical analysis of this study was performed by utilizing statistical software, Statistical Package for Social Sciences (SPSS) for Windows 17.0.

Table I Shows the distribution of study sample according to the type of CHD

	Frequency	Percent
AVSD	15	32
VSD	12	26
PDA	8	17
ASD	7	15
Complex	4	8
TOF	1	2
Total	47	100

Results

During the study period 109 infants with a diagnosis of Down's syndrome were seen. Infants were divided into three groups according to age. Less than one month 41(37.6%), between one month and six months 37(34%) and more than six months up to one year 31(28.4%). 56 infants were females and 53 were males. The study showed that 47(43.1%) infants had CHD while 62(56.9%) had no CHD. The commonest cardiac lesion was Atrioventricular septal defect (AVSD) followed by VSD, PDA and Secondum ASD Table 1, all lesions were single and there were no multiple lesions. The nutritional status of the study group showed that 46(42.2%) were normal, 62(56.9%) were under nourished and one (0.9%) was overweight.

Discussion

The incidence of CHD in DS infants in our population is 43% with AVSD the commonest lesion (32%), this is similar to other studies conducted in USA and Europe^{7,15} which showed CHD incidence of 43% and 44% respectively. A higher incidence was reported by Jaiyesemi & Baichoo¹⁶ from Oman showing a prevalence of 67% with the AVSD as the commonest lesion (32%) followed by ASD (29%), PDA (17%) and VSD (14%). In a previous study in Khartoum state including 96 children with Down syndrome (age 2 month-15 years), CHD was diagnosed in 37.5%, the commonest lesion were VSD (47%) followed by AVSD (22%), ASD (11%) and

TOF (5%).¹⁷ In another study from Sudan by, CHD was found in 73 out 80 Down's syndrome children followed in a cardiac centre, the commonest lesion were AVSD in 48% and VSD in 23%; this high figures could be explained by the fact that these patients were already followed in a cardiac centre.¹⁸

Table 2 Shows the Correlation between the nutritional status and presence of CHD

		Nutritional Status			Total	
		Under Nutrition	Normal	Obese		
CHD	Yes	Count	41	6	0	47
		% within CHD	87.20%	12.80%	0%	100.00%
		% within wt/length	66.10%	13.00%	0%	43.10%
	no	% of Total	37.60%	5.50%	0%	43.10%
		Count	21	40	1	62
		% within CHD	33.90%	64.50%	1.60%	100.00%
Total		% within wt/length	33.90%	87.00%	100.00%	56.90%
		% of Total	19.30%	36.70%	0.90%	56.90%
		Count	62	46	1	109
		% within CHD	56.90%	42.20%	0.90%	100.00%
		% within wt/length	100.00%	100.00%	100.00%	100.00%
		% of Total	56.90%	42.20%	0.90%	100.00%

(P < 0.001)

In our study 62(56.9%) infants were found to have under nutrition while 46(42.2%) were normal. 41(87.2%) of Down's syndrome with CHD were under nourished and this confirm that the presence of CHD was a major factor in affecting the nutritional status of infants with Down's syndrome (p value of < 0.001) Table 2. This is in keeping with several previous studies e.g., Crounk et al.¹³; Meguid et al.¹⁴; Tüysüz et al.¹⁹ & Bravo-Valenzuela et al.²⁰ All of these studies found that the affected age group is mainly the less than three years of age and the result of our study is keeping with the previous studies confirming that CHD has a detrimental factor in the growth of DS infants. To the best of our knowledge this is the first study done in Sudan looking at the impact of CHD in Down's syndrome infants on the nutritional status and the incidence of CHD in Down's syndrome in a younger and a homogenous population.

Conclusion

There is a high incidence of CHD among DS children, the commonest lesion is AVSD. The prevalence and pattern of CHD in DS is similar to studies from the western world. The presence of CHD is a detrimental factor for the nutritional status. All children suspected of having Down's syndrome should have a cardiac assessment including echocardiography as per international guidelines as soon as possible once the diagnosis of Down syndrome is suspected.

Acknowledgements

None.

Conflicts of interest

The authors declare that there are conflicts of interest.

Funding

None.

References

1. Torfs CP, Christianson RE. Anomalies in down syndrome individuals in a large population-based registry. *Am J Med Genet.* 1998;77(5):431–438.
2. Freeman SB, Taft LF, Dooley KJ, et al. Population-based study of congenital heart defects in down's syndrome. *Am J Med Genet.* 1998;80(3):213–217.
3. Vis JC, Duffels MG, Winter MM, et al. Down syndrome: a cardiovascular perspective. *J Intellect Disabil Res.* 2009;53(5):419–425.
4. Roizen NJ, Patterson D. Down's syndrome. *Lancet.* 2003;361(9365):1281–1289.
5. Rasmussen SA, Wong LY, Correa A, et al. Survival in infants with down syndrome, Metropolitan Atlanta, 1979–1998. *J Pediatr.* 2006;148(6):806–812.
6. American academy of pediatrics committee on genetics. Health supervision for children with down syndrome. *Pediatrics.* 2001;107(2):442–449.
7. Freeman SB, Bean LH, Allen EG, et al. Ethnicity, sex, and the incidence of congenital heart defects: a report from the National down syndrome Project. *Genet Med.* 2008;10(3):173–180.
8. Vida VL, Barnoya J, Larrazabal LA, et al. Castañeda AR congenital cardiac disease in children with down's syndrome in Guatemala. *Cardiol Young.* 2005;15(3):286–290.
9. Toledo C, Alembik Y, Aquirre Jaime A, et al. Growth curves of children with down syndrome. *Ann Genet.* 1999;42(2):81–90.
10. Myrelid A, Gustafsson J, Ollars B, et al. Growth charts for Down's syndrome from birth to 18 years of age. *Arch Dis Child.* 2002;87(2):97–103.
11. Al Husain M. Growth charts for children with down's syndrome in Saudi-Arabia: birth to 5 years. *Int J Clin Pract.* 2003;57(3):170–174.
12. Afifi HH, Aglan MS, Zaki ME, et al. Growth charts of down syndrome in Egypt: a study of 434 children 0–36 months of age. *Am J Med Genet.* 2012;158A(11):2647–2655.
13. Cronk C, Crocker AC, Pueschel SM, et al. Growth charts for children with down syndrome: 1 month to 18 years. *Pediatrics.* 1988;81(1):102–110.
14. Meguid NA, El-Kotoury AI, Abdel-Salam GM, et al. Growth charts of Egyptian children with down syndrome (0–36 months). *East Mediterr Health J.* 2004;10(1–2):106–115.
15. Weijerman ME, van Furth AM, van der Mooren MD, et al. Prevalence of congenital heart defects and persistent pulmonary hypertension of the neonate with down syndrome. *Eur J Pediatr.* 2010;169(10):1195–1199.
16. Jaiyesimi O, Baichoo V. Cardiovascular malformations in Omani Arab children with down's syndrome. *Cardiol Young.* 2007;17(2):166–171.
17. Nuha Sharaf El, Din El. Incidence and pattern of CHD in Sudanese children with down's syndrome. Clinical paediatric MD book of abstracts. (2nd edn), University of Khartoum post graduate medical studies board. 2004:202–203pp.
18. Ali SK. Cardiac abnormalities of Sudanese patients with down's syndrome and their short-term outcome. *Cardiovasc J Afr.* 2009;20(2):112–115.
19. Tüysüz B, Göknar NT, Oztürk B. Growth charts of Turkish children with down's syndrome. *Am J Med Genet.* 2012;158A(11):2656–2664.
20. Bravo-Valenzuela NJ, Passarelli ML, Coates MV, et al. Weight and height recovery in children with down syndrome and congenital heart disease. *Rev Bras Cir Cardiovasc.* 2011;26(1):61–68.