Original Article

Prevalence of congenital heart disease among Palestinian children born in the Gaza Strip

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Abstract *Objective:* This study was designed to estimate the birth prevalence of children with congenital heart disease born in the Gaza Strip during 2010 and to compare these with estimates from elsewhere. *Methods:* We reviewed the medical records of all children born in 2010 who were diagnosed, treated, and/or followed up in the four paediatric cardiology clinics in the Gaza Strip. Data were also obtained from El Makassed Hospital in East Jerusalem and from the Schneider Hospital, Wolfson Medical Center, and Tel HaShomer Hospital in Israel, where we had referred some of our patients for percutaneous or surgical treatment. *Results:* A total of 598 children with congenital heart disease were detected among the 59,757 children born alive in the Gaza Strip during 2010, yielding a birth incidence of 10 per 1000 live births. The most frequently occurring conditions were ventricular septal defects (28%), ostium secundum atrial septal defects (17%), patent ductus arteriosus (8.5%), and pulmonary valve abnormalities (8%). In this study, 7% of the children died. The actuarial survival at 6 months and 1 year of age was 94% and 93%, respectively, and remained stable over 18 months of follow-up. *Conclusion:* The birth incidence of congenital heart disease in the Gaza Strip in 2010 (10 per 1000) is higher than most estimates in Western Europe (8.2 per 1000 live births) and North America (6.9 per 1000 live births) but is similar to estimates from other parts of Asia (9.3 per 1000 live births).

CONGENITAL HEART DISEASE REFERS TO STRUCTURAL or functional heart disease that is present at birth.

It represents the most frequently occurring congenital anomaly in newborns.[1–6] In all, 28% of all major congenital anomalies consist of heart defects.[3] The estimate of eight per 1000 live births is generally accepted as the best approximation,[3–5] although large variations in the data occur across studies.[6] The causes of most congenital heart anomalies are unknown, but thought to be multifactorial and resulting from a combination of genetic predisposition and environmental stimuli. Advances in cardiovascular diagnostics and cardiothoracic surgery over the past century have led to increased survival of newborns with congenital heart disease,[7] and a dramatic increase in the population of school-aged and adult survivors of infant heart surgery. This has been accompanied by increased recognition of the long-term morbidities associated with congenital heart disease and has created a steadily growing population of patients with grown-up congenital heart disease. The prevalence of congenital heart disease in adults has been estimated to be four per 1000.[8] The aim of the present study was to describe the birth prevalence and treatments applied to Palestinian children with congenital heart disease who were born in the Gaza Strip in 2010, and to provide a cohort for long-term follow-up.

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Methods

Study population

The occupied Palestinian territory consists of two distinct areas: the West Bank, including East Jerusalem and the Gaza Strip. The Gaza Strip is about 41 km (25 miles) long, and between 6 and 12 km (4–7.5 miles) wide, with a total area of 360 sq km (140 square miles). It is home to a population of about 1.7 million people, most of them refugees or descendants of refugees. There are four clinics for paediatric cardiology in the Gaza Strip, one of them based at a tertiary care centre. The health-care system allows easy access to the paediatric cardiology clinics, and thus nearly all children with congenital heart disease in Gaza have been evaluated by the paediatric cardiology programmes at these four clinics. All patients registered in the databases of these programmes and born in Gaza in 2010 were eligible for inclusion in the present study. We only included cases proven by echocardiography in live births. We followed up these children for 18 months.

Measurements and definitions

Data obtained from the medical records included date of birth, postal code, gender, cardiac diagnoses, date and type of interventions, and date and cause of death. To determine whether a patient should be included in this study, we applied Mitchell's[9] definition of congenital heart disease to the cases under review: gross structural abnormalities of the heart or intrathoracic great vessels that are actually or potentially of functional significance. Patients with the following anomalies were excluded: isolated patent foramen ovale; rhythm disturbances without structural defects; isolated mild peripheral pulmonary stenosis; isolated innocent patent ductus arteriosus; patent ductus arteriosus in pre-term infants; hereditary disorders without cardiac consequences; and malpositioning of the heart without structural defects.

Congenital heart disease is often characterised by a combination of different heart lesions. To categorise patients according to their primary heart defect, we used a hierarchy of heart defects previously developed by the CONCOR (CONgenitale COR Vitia) project,[10] an initiative to form a national registry of patients with congenital heart disease in the Netherlands. Some modifications to the list of heart defects were made: univentricular heart was divided into hypoplastic left heart syndrome and univentricular physiology; truncus arteriosus was added; and Marfan syndrome and pulmonary arterial hypertension were removed from the list.[5] To calculate the birth prevalence of congenital heart disease and specific heart defects, we obtained the birth rate information in 2010 in the Gaza Strip from the database of the ministry of health.

Data analysis

The study was approved by the Human Research Ethics Committee. The overall incidence of congenital heart disease and the incidence of each type of defect were estimated and the type of surgery or catheterisation documented and tabulated.

Results

In 2010, 59,757 live births were registered in the Gaza Strip. In the four paediatric cardiology clinics, we recorded 598 children with congenital heart anomalies who were born alive in 2010. This yielded a birth prevalence of congenital heart disease of 10 per 1000 births (Table 1). The most prevalent heart defects in this sample were ventricular septal defects (28%); atrial septal defects type II (17%); patent ductus arteriosus (8.5%), and pulmonary valve abnormalities (8%) (Table 2). The gender distribution in the overall sample was 335 (56%) girls and 265 (44%) boys (Table 1). In all, 150 children (25%) had a cardiosurgical operation and 48 (8%) a catheter intervention. A total of 42 children (7%) died. Most of them died during the first 6 months of life. The actuarial survival of children with congenital heart disease 6 months and 1 year after birth was 94% and 93%, respectively. The reasons for death were no surgical intervention (n = 20); cardiac failure (n = 6); extracardiac reasons or multi-organ failure (n = 5); post-operative complications (n = 4); operative complications (n = 3); sudden death (n = 1); and pulmonary complications (n = 3) (Table 3).

Discussion

To the best of our knowledge, this is the first population-based investigation of the birth prevalence and treatment of children with congenital

Table 1. Total birth, gender and birth prevalence of congenital heart disease among infants born alive in the Gaza Strip in 2010.

Total births	Male	Male with congenital heart disease	Female	Female with congenital heart disease
59,757 (100%)	30,812 (51.5%)	265 (44%)	28,945 (48.5%)	335 (56%)

Table 2. Birth prevalence of each congenital heart anomaly.

Type of defect	Number	%	Treated patients Surgery either catheterisation
VSD	167	28	23
ASD	101	17	6
PDA	52	8.5	23
PS	48	8	18
TOF	36	6	23
TGA	30	5	23
COA	27	4.5	20
AVSD	27	4.5	17
TA	18	3	14
DORV	15	2.5	8
TAPVD	12	2	5
PA	15	2.5	12
DCM	12	2	0
AoS	9	1.5	2
HLHS	12	2	0
Shone C	6	1	2
Truncus arteriosus	6	1	1
Single V	3	0.5	1
Ebstein	2	0.33	0
Total	598	100	198

AoS = Aortic Stenosis; ASD = atrial septal defect; AVSD = atrioventricular septal defect; COA = coarctation of aorta; DCM = dilated cardiomyopathy; DORV = double outlet right ventricle; HLHS = Hypoplastic left heart syndrome; PA = pulmonary atresia; PDA = patent ductus arteriosus; PS = pulmonary stenosis; Single V = Single Ventrical; Shone C = Shone Complex; TA = Tricuspid atresia; TAPV = total anomalous pulmonary venous return; TGA = transposition of the great arteries; TOF = tetralogy of Fallot; VSD = ventricular septal defect.

Table 3. Causes of death.

Cause of death	Number
No surgical intervention	20
Cardiac failure	6
Extracardiac reasons or multi-organ failure	5
Post-operative complications	4
Operative complications	3
Pulmonary complications	3
Sudden death	1

Table 4. Birth prevalence of congenital heart anomalies.

Region	Prevalence per 1000
North America	6.9
Western Europe	8.2
Japan	9.3
Gaza Strip	10

heart disease in the occupied Palestinian territory. Its merit is that it includes the entire Gazan population of children with congenital heart disease. Another advantage of this population-based study is that it has identified a cohort for longitudinal follow-up studies to investigate the progress and outcomes of patient management. Our estimate of the birth prevalence of congenital heart anomalies of 10 per 1000 live births is higher than most estimates in Western Europe and in North America (8.2 and 6.9 per 1000 live births, respectively),[4,5,11] but close to Asian figures (9.3 per 1000 live births) (Table 4). Owing to the fact that babies who die perinatally in the Gaza Strip are not examined at autopsy, we may have underestimated the true number of serious heart defects. Indeed, as about 8% of heart defects are diagnosed after the age of 5 years,[12] the true estimate may be even higher.

The birth prevalence of individual heart defects found in our study is not entirely consistent with that of previous reports. Similar to other studies, ventricular septal defects, atrial septal defects, and pulmonary valve abnormalities have been the most frequently occurring heart defects,[5,13–21] but surprisingly a higher prevalence of major heart defects – hypoplastic left heart syndrome, 2 versus 1 per 10,000; pulmonary atresia, 2.5 versus 1 per 10,000; tricuspid atresia, 3 versus 0.5 per 10,000; and total anomalous pulmonary venous return, 2 versus 1 per 10,000 – were observed compared with the literature.[5,11] Obviously this calls for further genetic and environmental investigation. Ventricular septal defect was the most encountered heart defect in our study, accounting for 28% of the cases. The high prevalence of ventricular septal defect in our study and in other reports might be due to the fact that most children who present to paediatric cardiology units are still in the neonatal phase and do so because they show clinical signs and symptoms, for example heart murmur. The highest reported prevalence of ventricular septal defect in neonates,[22] of which close spontaneously between 1 and 10 months of life, can therefore be responsible for the high number of children with ventricular septal defect.[16] In all, 14% of our patients with ventricular septal defects were operated on to close the defect, which is relatively higher than in other reported studies.[5]

The low prevalence of left ventricular outflow tract obstructions – coarctation and aortic stenosis – contrasts sharply with European figures.[11] The same was observed in Japan, another Asian country, where aortic stenosis and coarctation of the aorta were reported in only 3.7% of all patients with congenital heart disease. Studies of ethnic influence on the pattern of congenital heart disease in the United Kingdom revealed a higher frequency of coarctation of the aorta in non-Asian (9%) compared

with Asian (3%) infants. This might be related to racial and genetic factors.

All cases of hypoplastic left heart syndrome were born between June and September, which seems to be very strange and unexplained and needs further confirmation of continuous registration.

Overall, 33% of our sample received an intervention during the course of the 18-month follow-up period. Catheter interventions were performed in 8% of the children in our series. The proportion of patients that underwent a cardiosurgical operation is in keeping with previous reports of patients with univentricular physiology; tetralogy of Fallot; transposition of the great arteries; atrioventricular septal defect; pulmonary valve abnormalities; and ventricular septal defects. The survival of newborns with congenital heart disease continues to improve. Although the overall life expectancy of these newborns is high, vulnerable subgroups do exist. Univentricular physiology, pulmonary atresia with ventricular septal defect, tetralogy of Fallot, and left ventricular outflow tract obstruction was associated with significantly higher mortality than other heart defects. All our patients with hypoplastic left heart syndrome left and expired without surgery.

Study limitations

The children were referred to paediatric cardiology when a clinical problem was detected. In the Gaza Strip, deliveries mainly occur in hospitals and a routine assessment of every newborn by a pediatrician within the first 7 days of life is common practice. Thus, heart murmurs are easily detected. However, systematic echocardiographical screening of the entire population is not done, and thus subclinical heart defects, such as subclinical ventricular septal defects, mild pulmonary stenosis, or small atrial septal defect, have been missed. Furthermore, we excluded isolated peripheral pulmonary stenosis, as well as patent ductus arteriosus and patent foramen ovale, which would not require closure. This may have affected the birth prevalence calculation in the present study. The higher birth prevalence of congenital heart disease compared with published estimates from elsewhere may be attributable to one or more of several factors, including low socio-economic status, multigravidity, the environment, genetic and ethnic factors, and high consanguinity rates, but we do not have data to explore these possibilities.

Conclusion

- This population-based study conducted in the Gaza Strip yielded a birth prevalence of congenital heart disease of 10 per 1000 live births.

- The birth prevalence of congenital heart disease and the prevalence of specific heart defects are not consistent with estimates from elsewhere.
- What is surprising to us is the higher prevalence of major heart defects, compared with other reported studies and the literature, such as hypoplastic left heart syndrome, pulmonary atresia, tricuspid atresia, total anomalous pulmonary venous return. This should be compared with other surrounding countries in order to find possible genetic or social explanations.
- Survival is nevertheless good, suggesting an ongoing improvement of life expectancy. Hence, the prevalence of congenital heart disease in the community will probably continue to increase.
- The findings of this study are not only relevant for clinicians and hospital administrators, but are also relevant for public health. Furthermore, this study can serve as a basis for the longitudinal follow-up of this cohort of patients, with the long-term aim of exploring the progress and outcomes of patient management.

Conflicts of Interest

None.

References

1. Dastgiri S, Stone DH, Le-Ha C, Gilmour WH. Prevalence and secular trend of congenital anomalies in Glasgow, UK. Arch Dis Child 2002; 86: 257–263.
2. Tagliabue G, Tessandori R, Caramaschi F et al. Descriptive epidemiology of selected birth defects, areas of Lombardy, Italy, 1999. Popul Health Metr 2007; 5: 4.
3. Dolk H, Loane M, Garne E. European Surveillance of Congenital Anomalies (EUROCAT) Working Group. Congenital heart defects in Europe: prevalence and perinatal mortality, 2000 to 2005. Circulation 2011; 123: 841–849.
4. Bernier PL, Stefanescu A, Samoukovic G, Tchervenkov CI. The challenge of congenital heart disease worldwide: epidemiologic and demographic facts. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2010; 13: 26–34.
5. Moons P, Sluysmans T, De Wolf D, et al. Congenital heart disease in 111 225 births in Belgium: birth prevalence, treatment and survival in the 21st century. Acta Paediatr 2009; 98: 472–477.

6. Hoffman JI, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol 2002; 39: 1890–1900.

7. Report of the British Cardiac Society Working Party. Grown-up congenital heart (GUCH) disease: current needs and provision of service for adolescents and adults with congenital heart disease in the UK. Heart 2002; 88 (Suppl 1): i1–i14.

8. Khairy P, Ionescu-Ittu R, Mackie AS, Abrahamowicz M, Pilote L, Marelli AJ. Changing mortality in congenital heart disease. J Am Coll Cardiol 2010; 56: 1149–1157.

9. Mitchell SC, Korones SB, Berendes HW. Congenital heart disease in 56,109 births. Incidence and natural history. Circulation 1971; 43: 323–332.

10. van der Velde ET, Vriend JW, Mannens MM, Uiterwaal CS, Brand R, Mulder BJ. CONCOR, an initiative towards a national registry and DNA-bank of patients with congenital heart disease in the Netherlands: rationale, design, and first results. Eur J Epidemiol 2005; 20: 549–557.

11. van der Linde D, Konings EE, Slager MA, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. J Am Coll Cardiol 2011; 58: 2241–2247.

12. Wren C, O'Sullivan JJ. Survival with congenital heart disease and need for follow up in adult life. Heart 2001; 85: 438–443.

13. Bosi G, Garani G, Scorrano M, Calzolari E. Temporal variability in birth prevalence of congenital heart defects as recorded by a general birth defects registry. J Pediatr 2003; 142: 690–698.

14. Calzolari E, Garani G, Cocchi G, et al. Congenital heart defects: 15 years of experience of the Emilia-Romagna Registry (Italy). Eur J Epidemiol 2003; 18: 773–780.

15. Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. Circulation 2007; 115: 163–172.

16. Hoffman JI, Kaplan S, Liberthson RR. Prevalence of congenital heart disease. Am Heart J 2004; 147: 425–439.

17. Forrester MB, Merz RD. Descriptive epidemiology of selected congenital heart defects, Hawaii, 1986–1999. Paediatr Perinat Epidemiol 2004; 18: 415–424.

18. Garne E. Congenital heart defects – occurrence, surgery and prognosis in a Danish County. Scand Cardiovasc J 2004; 38: 357–362.

19. Grech V. Spectrum of congenital heart disease in Malta. An excess of lesions causing right ventricular outflow tract obstruction in a population-base study. Eur Heart J 1998; 19: 521–525.

20. Robida A, Folger GM, Hajar HA. Incidence of congenital heart disease in Qatari children. Int J Cardiol 1997; 60: 19–22.

21. Samanek M, Voriskova M. Congenital heart disease among 815,569 children born between 1980 and 1990 and their 15-year survival: a prospective Bohemia survival study. Pediatr Cardiol 1999; 20: 411–417.

22. Roguin N, Du ZD, Barak M, Nasser N, Hershkowitz S, Milgram E. High prevalence of muscular ventricular septal defect in neonates. J Am Coll Cardiol 1995; 26: 1545–1548.