Congenital malformations at a referral hospital in Gorgan, Islamic Republic of Iran

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التشوُّهات الخلقية في مستشفى للإحالة في جرجان، بجمهورية إيران الإسلامية

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الخلاصة: تم في هذه الدراسة تسجيل معدل التشوُّهات الخلقية في 10 000 ولادة في مستشفى للإحالة في مدينة جرجان الإيرانية، في المدة 1998 – 1999. وبيَّنت الدراسة أن معدل الوقوع الإجمالي لهذه التشوُّهات قد بَلَغَ 1.01٪ (1.19٪ في الذكور، 0.76٪ في الإناث). وكانت شذوذات الجهاز العضلي الهيكلي هي الأكثر وقوعاً (0.38٪)، تليها شذوذات الجهاز العصبي المركزي (0.28٪)، ثم شذوذات الجهاز البولي التناسلي (0.25٪). وكان معدل وقوع التشوُّهات الخلقية في الفئات الأَثنَيَّة مختلفاً من فئة إلى أخرى، إذْ بَلَغَ 0.85٪ بين الفُرْس، و1.45٪ بين التركمان، و1.70٪ بين السستانيِّين. وتدل الدراسة على أن الجنس والأصل الأَثنَي من العوامل المؤثِّرة في معدل وقوع التشوُّهات الخلقية في هذه المنطقة.

ABSTRACT This study recorded the rate of congenital malformations in 10 000 births at a referral hospital in Gorgan, Islamic Republic of Iran in 1998–99. The overall incidence of congenital malformations was 1.01% (1.19% in males and 0.76% in females). Anomalies of the musculoskeletal system had the highest incidence (0.38%), followed by central nervous system (0.28%) and genitourinary system (0.25%). The incidence of congenital malformations in different ethnic groups was 0.85%, 1.45% and 1.70% in native Fars, Turkman and Sistani groups respectively. Sex and ethnic background are factors in the rate of congenital malformations in this area.

Les malformations congénitales dans un hôpital de recours à Gorgan (République islamique d'Iran)

RÉSUMÉ Cette étude portait sur le taux de malformations congénitales pour 10 000 naissances recensé dans un hôpital de recours à Gorgan (République islamique d'Iran) en 1998-1999. L'incidence globale des malformations congénitales était de 1,01 % (1,19 % chez les garçons et 0,76 % chez les filles). L'incidence la plus élevée était celle des anomalies du système ostéo-articulaire et des muscles (0,38 %), suivies par les anomalies du système nerveux central (0,28 %) et de l'appareil génito-urinaire (0,25 %). L'incidence des malformations congénitales dans différents groupes ethniques s'élevait à 0,85 %, 1,45 % et 1,70 % pour les groupes autochtones Fars, Turkman et Sistani respectivement. Le sexe et l'origine ethnique sont des facteurs qui influencent le taux de malformations congénitales dans cette région.

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Introduction

The leading causes of infant morbidity and mortality in poorer countries are malnutrition and infections [*1*], whereas in developed countries they are cancer, accidents and congenital malformations [*2,3*]. In the Islamic Republic of Iran, an effective and comprehensive programme of vaccination has been implemented, childhood malnutrition has been eliminated and serious childhood infections are disappearing. Therefore, congenital malformations will begin to emerge as one of the major childhood health problems. Treatment and rehabilitation of children with congenital malformations is costly, and complete recovery is usually impossible [*4,5*]. Considerable variations in the frequency of congenital malformations in different populations have been reported, from 1.07% in Japan [*6*] to 4.3% in Taiwan [*7*].

Surveys on congenital malformations in the Islamic Republic of Iran have been carried out in Tehran [*8*], Arak [*9*] and Hamadan [*10*] cities in the central area. There may be regional variations in the pattern of congenital anomalies and to the best of our knowledge, a similar study has not been conducted among newborns in Golestan province in the north of the country. The present study was carried out to record the pattern of congenital malformations in this area of the Islamic Republic of Iran.

Methods

This was a descriptive, cross-sectional study of newborn and stillborn babies delivered in Dezyani hospital during a 20-month period, January 1998 to August 1999. Dezyani hospital serves as a referral centre for obstetrics and gynaecology and prenatal intensive care for all other hospitals and clinics in Golestan province in the south-east Caspian Sea area. The region has a population of about 1.5 million and covers an area of about 20 460 square kilometres. Dezyani hospital is one of 17 hospitals, which covers more than 500 primary health centres in this region. As a result, patients in these catchment areas requiring transfer for specialized obstetric and gynaecology investigations and treatments must be referred to Dezyani hospital.

All live and stillborn newborns delivered in this hospital during the investigation were examined and screened for congenital malformations by a paediatrician. The charts of newborns with congenital malformations were subsequently extracted for detailed study. Variables recorded included demographic data: the date of admission, sex, ethnicity and maternal age. Ethnicity was classified into 3 groups: native Fars, Turkman and Sistani. The native Fars are the original and main inhabitants of the region. The Turkman immigrated from the Middle East from 250 years ago and the rate of inter-racial marriage among the Turkman is nearly 100%. The Sistani originated from the Iran–Pakistan–Afghanistan border from half a century ago. Clinical information recorded included: treatment and outcome at discharge (transferred or dead). No autopsy examinations were performed on those newborns that died after birth.

The data were analysed to highlight the pattern and the relative importance of the different types of congenital malformations among the newborns in this region.

The types of birth defects were classified by the diagnostic standardization of congenital malformation from the *International classification of diseases (ICD-10)* codes. The data were analysed by *SPSS*, version 10. The rate of malformed newborns and the rate of malformations were compared with statistical *t*-test and chi-

squared tests. The level of significance was $P < 0.05$.

Results

During the 20-month period, 10 000 newborns were delivered and admitted to Dezyani hospital: 5048 males and 4880 females. There were 7790 native Fars, 1035 Turkman and 1175 Sistani.

Out of these, 101 newborns were diagnosed with congenital malformations, giving an incidence in this sample of 1.01%. The male to female ratio was 1:1.6 (60 males, 37 females, 4 with ambiguous genitalia) (Table 1). The rate of congenital malformation was significantly different between male and female newborns (1.19% and 0.76%, respectively; $P < 0.05$).

Of the 101, 66 were native Fars, 15 Turkman and 20 Sistani. Thus, the incidence of congenital malformations was 0.85%, 1.45% and 1.70% in native Fars, Turkman and Sistani groups respectively ($P < 0.05$).

Table 2 shows the *ICD-10* classification of the different types of congenital malformation. Some newborns had a multiplicity of malformations, so that the total number of congenital malformations exceeded the number of affected newborns. Altogether, 156 anomalies were documented in 101 newborns.

The musculoskeletal system was the most affected, involving 38 out of 101 patients. Among this group, the most frequent lesions were clubfoot, polydactyly and meromelia. The central nervous system came second in frequency, involving 28 newborns. Meningomyelocele, followed by anencephaly, meningocele and hydrocephaly were the most prominent central nervous system lesions. Anomalies of the genitourinary system were the next highest group, involving 25 out of 101 patients. The most common anomaly was hypospadias. Digestive system problems involved 15 patients, and imperforate anus was the most common lesion detected.

Twelve out of 101 (11.9%) patients with congenital malformations died, although the total deaths among the 10 000 newborns was 68 (0.68%) (Table 3). The death rate was relatively higher in patients with malformations of the central nervous system.

Discussion

In the present study, the overall incidence of congenital malformations among newborns was 1.01%. This result is very similar to previous data from the Islamic Republic of Iran: 1.04% in Arak in the central area [*9*] and 1.18% in Tehran [*11*]. However, it is lower than that reported in Tehran in 1986 (3.5%) [*8*] and lower than

Table 1 **Incidence of congenital malformations (CM) by sex**

Sex[a]	No. newborns delivered	No. with CM	%	Relative risk	95% CI
Male	5048	60	1.19	1.21	1.04–1.42
Female	4880	37	0.76		

CI = confidence interval.
[a]Four newborns had ambiguous genitalia.

Table 2 **All births with congenital malformations (CM) by system according to the international Classification of Diseases (*ICD-10*)**

Malformations/system	No. of CM	%	Rate per 10 000 births
Musculoskeletal system			47
Clubfoot	15	9.6	
Polydactylia	14	9.0	
Meromelia	8	5.1	
Achondroplasia	4	2.6	
Syndactyly	2	1.3	
Lobster claw	1	0.6	
Others	3	1.9	
Central nervous system			36
Meningomyelocele	10	6.4	
Meningocele	8	5.1	
Hydrocephaly	8	5.1	
Anencephaly	8	5.1	
Microcephaly	2	1.3	
Genitourinary system			26
Hypospadias	18	11.5	
Ambiguous genitalia, hermaphroditism	4	2.6	
Epispadias	3	1.9	
Kidney disease	1	0.6	
Digestive system			17
Imperforate anus	13	8.3	
Umbilical hernia	2	1.3	
Omphalocele	2	1.3	
Eye, ear, face and neck	6	3.8	6
Oral/cleft lip and palate			14
Cleft lip and cleft palate	6	3.8	
Cleft lip	4	2.6	
Cleft palate	4	2.6	
Chromosomal anomalies			
Down syndrome	6	3.8	6
Other anomalies			4
Ichthyosis (collodion baby and harlequin fetus)	2	1.3	
Thoracopagus twins	1	0.6	
Tumour	1	0.6	
Total	156	100.0	

reports from other populations (1.27%) in World Health Organization centres in 16 countries [*12*]. Other studies showed different prevalence figures: in India (1.28%)

Table 3 **Mortality among normal and congenital malformed (CM) newborns at birth**

Outcome	No. of normal newborns	No. of newborns with CM	Total
Live births	9839	89	9928
Stillbirths	56	12	68
All births	9895	101	9996
	χ^2 = 189, ***P*** < 0.001, OR = 23.7		

OR = odds ratio.

[*13*], in Spain (2.02%) [*14*], in Bahrain (2.7%) [*15*], in Egypt (3.17%) [*16*], in the Libyan Arab Jamahiriya (0.93%) [*17*] and in Atlanta, USA (3.1%) [*18*]. Table 4 shows the incidence of birth defects in parts of the Islamic Republic of Iran and other countries. These variations between different studies could be explained by the effect of different racial, ethnic and social factors in various parts of the world or different geographical, nutritional and socioeconomic factors. Other explanations for these variations in birth defect incidence are the type of sample and the criteria for diagnosis.

The rate of malformations in male newborns is nearly twice that of females. This result is the same as a report from Arak [*9*] and other reports from different countries [*12*].

The commonest system involved was the musculoskeletal system, which agrees with reports from other parts of the Islamic

Table 4 **Incidence of congenital malformations (CM) in Gorgan, other regions of the Islamic Republic of Iran and selected countries**

Location/reference	Rate of CM per 1000 Live births	All births
Benghazi, Libyan Arab Jamahiriya [***17***]	70	9.3
Tehran, Islamic Republic of Iran [***8***]	35	–
Atlanta, United States of America [***18***]	31	–
Giza, Egypt [***16***]	–	31.7
Bahrain [***15***]	27	–
Kabul, Afghanistan [***34***]	24	–
Saudi Arabia [***35***]	22.7	–
Beirut, Lebanon [***36***]	–	16.5
Spain [***14***]	20.3	–
Western area, China [***33***]	–	15.4
Singapore [***37***]	15.1	–
Tehran, Islamic Republic of Iran [***12***]	11.8	–
Alexandria, Egypt [***16***]	–	11.6
Maharashtra, India [***13***]	10.8	12.8
Arak, Islamic Republic of Iran [***9***]	10.4	–
Gorgan, Islamic Republic of Iran (present study)	–	10.1

Republic of Iran [*8,9*] and other countries [*12,13*].

Table 5 compares the incidence of the difference type of congenital malformations in our study with other studies in the Islamic Republic of Iran and elsewhere. The most common musculoskeletal anomaly was clubfoot (1.5 per 1000), the same as a study in Tehran (1.5 per 1000) and in Arak (1.4 per 1000) [*9,11*], but lower than another report in Tehran of 2.91 per 1000 [*8*]. It is noticeable that in our study 10 out of 15 newborns with clubfoot were female; this contrasts with other investigations [*8,11*]. The rate of neural tube defects is higher than other studies in Tehran, Islamic Republic of Iran [*8,11*], and other surveys in different countries [*13,17,19–22*], but lower than China [*23*] and the north-west Islamic Republic of Iran [*24*].

The most common malformation of the central nervous system was spina bifida cystica (meningomyelocele and meningocele), the same as one report from Tehran [*11*]. The rate of spina bifida (1.8 per 1000) was lower than in Britain and USA [*25*] and higher than in India with 0.34 per 1000 and 0.38 per 1000 in Tehran and 0.7 per 1000 in north-west of the Islamic Republic of Iran [*8,13,24*]. The rate of anencephaly was 0.8 per 1000, which was lower than other reports from India and the Islamic Republic of Iran [*11,13,24*], higher than the survey in Tehran [*8*] and in USA, Brazil and Africa [*25*]. As in other reports [*11,25*], spina bifida in our study was more common in females.

The rate of cleft palate was 0.4 per 1000, lower than one report in Tehran [*11*]. The rate of cleft palate with or without cleft lip was 1.4 per 1000, nearly similar to studies in England, lower than in the USA [*25*] and higher than in Tehran [*8,11*] and elsewhere [*12,13,26*].

Table 5 **Comparison of different type of congenital malformations (CM) in Gorgan with other studies**

Location	Rate of CM per 1000 births						
	Club-foot	Cystic spina bifida	Anence-phaly	Cleft palate + or – cleft lip	Imper-forate anus	Hypospa-diasis	Down syn-drome
Gorgan, Islamic Republic of Iran (present study)	1.5	1.8	0.8	1.4	1.3	1.8	0.6
Arak, Islamic Republic of Iran [***9***]	1.4	1.2	0.4	1.6	–	–	–
Tehran, Islamic Republic of Iran 1991 [***11***]	1.5	1.8	1.0	1.5	0.1	–	1.3
Tehran, Islamic Republic of Iran 1986 [***11***]	2.90	0.92	0.08	1.61	0.38	4.45	1.23
Britain [***3,38***]	1.2	3.4	3.9	1.3	0.4	1.9	1.3
United States of America [***3, 39, 40,41***]	–	0.8	0.5	1.6	0.43	3.9	0.7
Maharashtra, India [***13***]	1.04	0.34	0.69	1.04	–	0.34	0.69
Giza, Egypt [***16***]	–	0.66	–	1.66	0.33	2.33	1.33

The most common genitourinary malformation was hypospadiasis and the rate was lower than in other parts of the Islamic Republic of Iran [*8,27*] and higher than in the south-west of the country [*28*] and India [*13*]. The rate of imperforate anus was 1.3 per 1000, higher than other studies in the Islamic Republic of Iran [*8,11*], in Saudi Arabia 0.6 per 1000 [*29*] and in Denmark 0.38 per 1000 [*30*].

In our study, the rate of Down syndrome was 0.6 per 1000, which is lower than reports from Tehran [*8,11*], USA, Brazil, England and India [*25,31*], but higher than in central Africa [*25*].

The rate of congenital malformation varies in different races. Previous studies in the USA indicated that the rate of neural tube defects and spina bifida have some differences across various racial and ethnic groups [*21,32*]. We need more investigations in our region in order to determine the role of racial and ethnic factors.

Infant morbidity and mortality due to congenital malformation in the Islamic Republic of Iran was lower than China and higher than the USA [*33*], but the finding that central nervous system anomalies were the highest cause of death is similar to other research [*33*]. The difference between the incidence of types of congenital malformation in different parts of this country may be due not only to genetic background but also to geographical, nutritional and socioeconomic differences.

More in-depth analytic research is needed to determine the possible genetic, sociodemographic and socioenvironmental factors underlying the various types of congenital malformation encountered in this area.

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