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ORIGINAL ARTICLE

Gastrointestinal malformations in Gorgan, North of Iran: epidemiology and associated malformations

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Abstract The aim of this prospective study was to evaluate the prevalence and pattern of gastrointestinal malformations (GIM) among Iranian newborns in Gorgan, North of Iran. From 1998 through 2003, 37,951 live births in Dezyani hospital in Gorgan, North of Iran, were screened for gastrointestinal malformations. Clinical and demographic factors of diagnosed cases were recorded in a pre-designed questionnaire for analysis; sex, ethnicity, type of GIM and associated anomalies. The overall prevalence rate of gastrointestinal malformations was 10 per 10,000 births. The imperforate anus (5 per 10,000) was the commonest birth defect in gastrointestinal tract. The prevalence rate of GIM was 8.2 per 10,000 in males and 10.7 per 10,000 in females. According to the parental ethnicity, the prevalence rates of GIM were 6.7, 15.8 and 17.6 per 10,000 in Fars, Turkman, and Sistani, respectively. There were eight cases (21%) with associated anomalies. The prevalence rate of GIM in North of Iran is not similar to the previous studies in Iran and Middle East and ethnic background may be a causative factor in the rate of GIM in this area.

Keywords Gastrointestinal malformations · Prevalence · Ethnicity

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Introduction

So far, a comprehensive report focusing specifically on congenital anomalies of gastrointestinal tract, in Iran, has not yet been documented. In some previous general surveys performed in a short period on congenital malformations in Tehran and Gorgan, North of Iran, the gastrointestinal system was found to be one of the important systems affected [1–3].

The majority of infants need surgery in the neonatal period in order to survive [4]. A high proportion of infants with gastrointestinal malformations (GIM) have associated malformations [5] and mortality rate among these infants is higher than those infants with isolated GIM [6, 7].

Although there were some reports on congenital anomalies of gastrointestinal tract in the world [4, 8–10] there is no focused study in Iran and North of Iran. Therefore, this study was done to determine the prevalence pattern of congenital lesions, associated anomalies and ethnicity among Iranian newborns in Gorgan, North of Iran; South-East of Caspian Sea border.

Patients and methods

This investigation was undertaken on 37,951 live births, to identify all newborns with gastrointestinal malformations, born between January 1, 1998 and December 31, 2003, in Dezyani—a teaching hospital and a referral center which is the main site for about 70% of deliveries in Gorgan—a capital city in Golestan province in the north of Iran. All live newborns delivered in this hospital during the investigation were examined and

screened for GIM including atresia (Esophageal and Anal atresia), Gastroshisis, Omphallocell, Umbelical hernia and diaphragmatic hernia.

The charts of newborns with GIM were subsequently extracted for detailed study. Variables recorded included: the date of birth, sex, type of malformations, ethnicity of parents and the presence of other congenital malformations. The total number of live figures of every year was recorded.

According to their ethnicity, three ethnic groups (Native Fars, Turkman and Sistani) who are residing in this region were considered. The native Fars group is the predominant inhabitant of the region. The Turkmans are those who have emigrated from the central Asia since 250 years ago. The Sistani group emigrated from Iran–Pakistan–Afghanistan border (Sistan and Baluchestan province in South-East of Iran) since half a century ago.

Statistical methods

Descriptive data are presented as percentages. Descriptive statistics was calculated for GIM prevalence per 10,000 live births. The prevalence of GIM is calculated as follows:

$$\text{Annual rate} = \frac{\text{The GIM cases}}{\text{The total live birth}}.$$

Confidence interval (CI 95%) for prevalence was calculated by binomial exact methods. Chi-square for trend test was used to assess trend over time. STATA 8/SE statistical package was used for statistical analysis.

Results

A total of 38 newborns with 40 GIM, were registered during the study period, giving a prevalence of 10 cases (CI 95% 7.1–13.7) per 10,000 births and 10.54 (CI 95% 7.5–14.4) malformations per 10,000 births. In two cases, there were more than one GIM and both of them included anal Atresia.

The prevalence of GIM was 11.39 per 10,000 (7.1–17.1) and 8.71 per 10,000 (5–14.1) during the two periods 1998–2000 and 2001–2003, respectively (Fig. 1).

The prevalence of GIM in males and females was 8.3 per 10,000 and 10.8 per 10,000, respectively. Risk of GIM in female was 1.3 times more than male (odds ratio = 1.3) but this difference was not significant (CI 95% 0.7–2.5) (Table 1).

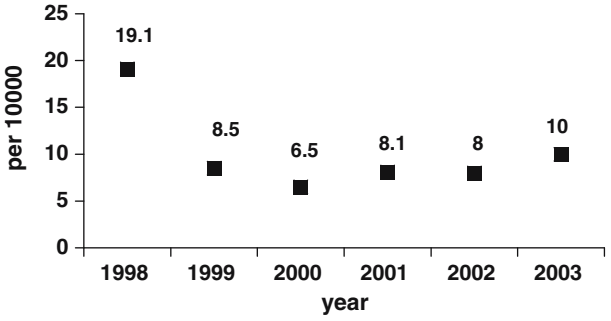

Fig. 1 Prevalence of gastrointestinal malformations in North of Iran 1998–2003. Rate per 10,000 birth trend time

Anal Atresia with prevalence rate of 5 per 10,000 was the most common GIM malformations. This prevalence rate was 3.1 (1.1–6.7) per 10,000 and 6.46 (3.3–11.3) per 10,000 in males and females, respectively. The prevalence rate of gastrointestinal malformations is depicted in Table 1. Risk of anal atresia in females was 2.1 times more than males (odds ratio = 2.1) but this difference was not significant (CI 95% 0.7–6.7) (Chi-square = 2.3, $P = 0.3$).

The prevalence of GIM was 6.7 per 10,000 births in the native Fars community, 15.8 per 10,000 in the Turkmans and 17.6 per 10,000 in Sistanis. Risk of GIM in Turkman and Sistani ethnicity groups were 2.4 (CI 95% for odds ratio 1–5.6) and 2.6 (CI 95% for odds ratio 1.2–5.8) times more than native Fars population. Twenty-one percent of the newborns with GIM had associated major anomalies including neural tube defects (two cases), Limb defects (four cases), cleft palate (one case) and epispadiases (one case).

Discussion

This investigation was undertaken to establish the prevalence rate of newborns with gastrointestinal malformations in Gorgan, in North of Iran. This study

Table 1 Total number of malformations in newborns with GIM, prevalence per 10,000 and proportion of cases with isolated gastrointestinal malformation

	Number	Prevalence per1,0000	Proportion with isolated GM (%)
Esophageal atresia	4	1	4 (100)
Anal atresia	19	5	12 (63)
Diaphragmatic hernia	4	9	4 (100)
Omphalocel	7	1.84	6 (86)
Gastroshisis	4	9	4 (100)
Umbelical hernia	2	0.53	2 (100)

was limited to newborns with defects that were clinically symptomatic during the newborn age bracket. It undoubtedly misses out some newborns whose congenital lesions, though present at birth, were silent or could have been misdiagnosed during the neonatal period.

By considering all these factors, our figures on birth prevalence are very likely to be an underestimation of the real situation. Besides, since Dezyani hospital is the only referral center in this region, it can be assumed that this center had attracted the bulk of Iranian newborns born with GIM anomalies in this area. Therefore, it is hoped that the results of this study demonstrate a clue, regarding the pattern of congenital gastrointestinal tract lesions in this area and perhaps in the entire Iran.

In this report, the prevalence of GIM was 10 cases per 10,000 births. Review of the literature did not reveal any investigations that reported the prevalence of GIM in separate long time study focused on GIM in Iranian population.

There have been some studies on congenital malformations that pointed out on GIM [1–3] that these investigations have been done in short period and limited population.

The results of the present investigation are higher than Farhouds' study [1] in Tehran with a prevalence rate of 6.9 per 10,000, but lower than another study in Tehran [2] with a prevalence rate of 39 per 10,000 and our previous study with prevalence rate of 17 per 10,000 [3].

In addition, our results are lower than a similar study in Saudi Arabia, with prevalence rate of 13 per 10,000 live births [8] and higher than other two studies in Egypt (6.6 per 10,000) and Bahrain (9 per 10,000) [11, 12]. Also, in a study in Denmark [4], the prevalence rate of GIM was 15.3 per 10,000 in all births. Prevalence rate of GIM according to the types of malformations as reported in literature is depicted in Table 2.

The difference between the results of different investigations in various parts of the world may be due to racial/ethnic, geographic, nutritional differences and also may be owing to difference in some place selections, data sources including live birth, still birth, abortion; classification and definition of GIM type and design of the study.

In this study, the prevalence rate of Atresia (esophageal and anal Atresia) was 6 per 10,000, which is lower than other studies in Saudi Arabia (12 per 10,000), in Denmark with 9.1 per 10,000 and in Europe with 7.6 per 10,000 [4, 5, 8].

The prevalence rate of anal Atresia was 5 per 10,000, which is lower than Asindi's study (6.1 per 10,000) in Saudi Arabia [8] and higher than other studies in Iran and in Denmark with 3.8 per 10,000 [1, 4] and in European countries with 4.05 per 10,000 [9].

The birth prevalence of anorectal malformation (5 per 10,000) observed in our region is nearly 2.5 times higher than the global figure with prevalence rate of 1:5,000 live births [13].

Other studies have shown a prevalence rate of esophageal Atresia of 4.1 per 10,000 [4], 2.8 per 10,000 [14], and 3 per 10,000 [8]. All these prevalence rates are higher than that found in our study. But the abovementioned difference may be due to regional properties such as Saudi Arabia and design of study such as in Denmark that included data from induced abortions.

In this study, the prevalence rate of diaphragmatic hernia, omphalocell and gastrochisis were, 1.05, 1.85 and 1.05 per 10,000, respectively. Other studies have shown a prevalence of diaphragmatic hernia of 2.4 per 10,000 [15], 2.1 per 10,000 [16], omphalocell 2.5 per 10,000 [17], 3 per 10,000 [4] and gastoschisis with 0.9 per 10,000 [17] and 1.3 per 10,000 [4].

The cause of low prevalence rate in our study in comparison with other studies [15–17] is due to sample setting, because these studies include data from live, stillbirth and induced abortions, but in our study only live births were considered.

Table 2 prevalence rate of GIM according to the types as reported in literature

	Location	Time span of study	Esophageal atresia	Anal atresia	Omphalocel	Gastroshisis	Diaphragmatic hernia	Total
Asindi et al. [8]	Saudi Arabia	1990–1995	3.28	6.1	–	–	–	13
Farhud et al. [1]	Tehran Iran	1969–1977	1.53	3.8	–	–	0.8	6.9
Garne et al. [4]	Funen country Denmark	1980–1993	4.1	3.8	3.0	1.3	2.7	15.3
Galzolari et al. [17]	European Country	1980–1990	–	–	2.5	0.9	–	–
Cuschieri [9]	Europe	1980–1994	–	4.05	–	–	–	–
Present study	Gorgan, Iran	1998–2003	1.05	5.01	1.85	1.05	1.05	10

Females are more affected than males. The female preponderance is seen in this survey in contrast with other researches [8, 18–20].

In this study, associated anomalies were seen in 21% of cases, but this rate was 35% in Garne's study in Denmark [4] and 22.7% in Saudi Arabia [8].

In present study, the commonest associated anomalies were limb anomalies, but in Garne's study in Denmark and Asindi's study in Saudi Arabia, it was cardiac malformations and Down's syndrome, respectively [4, 8]. Furthermore, another study was done in European countries on anorectal anomalies a long with other anomalies or as part of syndrome indicated almost 15% of cases that were chromosomal, monogenic or teratogenic syndromes, while the rest were of unknown causes including sequences (9.3%), VACTERL associations (15.4%) and multiple congentital anomalies (MCA) (60.2%) [10].

According to the ethnic factor the prevalence rate of GIM in Sistani group was higher than the other groups in this area. Although, a study in Hawaii did not find any relation between Anal atresia and ethnicity [21], but in other studies on the other congential malformations, it is shown that the ethnic factor could be an effective one in prevalence rate of malformations [3, 22–25].

High prevalence rate reported in Sistani ethnic group could also be not only due to ethnicity but also to the cultural habits or practices, linked to these particular ethnic groups. While, according to Asindi's findings in Arab population, the high prevalence of anorectal malformations in Saudi Arabia is due to inbreeding [8].

These differences in GIM risk between racial/ethnic groups within a given population may be due to difference in genetic factor or environmental risk factor associated with birth defects.

Conclusion

The present study showed the prevalence rate of GIM among Iranian newborns in Gorgan, in north of Iran, for the first time. These findings will assist in establishing the basis for a database in this area, but for determining the difference rate between ethnic groups and for surveying the etiological factors, further studies are required.

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