

Geographical distribution of congenital heart defects in Saudi Arabia

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BACKGROUND: Congenital heart defects (CHD), which are caused by abnormalities early in fetal life, encompass over 50 diagnoses. Since the detailed etiology is unknown, the geographical distribution of defects might suggest likely risk factors.

METHODS: The geographical distribution of 5 865 Saudi Arabian nationals with CHD was studied by cross-matching their residential provinces and towns with a geographical information system provided by the General Directorate for Military Survey. Population data were obtained from the 1413H census.

RESULTS: CHD cases were mostly distributed across the provinces in proportion to their total population but due to their size and inhomogeneity, province-based thematic maps were found to be misleading. City-based maps were preferable and showed similar geographic distributions for cases registered in successive years. Thematic maps of the distribution of the CHD burden highlighted the southwestern provinces, near the border with Yemen, and the northeast section of the Eastern Province.

CONCLUSIONS: Patterns of disease in Saudi Arabia are best studied at the level of individual towns and villages. The CHD registry has already attained good national coverage and can therefore support nationwide epidemiological studies. Southwestern Saudi Arabia and the northern part of the Eastern Province appear to exhibit a higher burden of CHD.

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Congenital heart defects (CHD) can encompass over 50 diagnoses. According to Mitchell,¹ a CHD can be defined as “a gross structural abnormality of the heart or intrathoracic great vessels that is actually or potentially of functional significance.” The majority of CHD cases are probably due to a genetic predisposition coupled with exposure to a teratogen² (either endogenous³ or exogenous⁴). However, the etiology is still largely unknown and the role of environmental factors is difficult to establish, given the rarity of the disease and the fact that most structural defects occur inter-utero in the first trimester of pregnancy. Establishing the geographical distribution of CHD patients would provide a convenient platform for the exploration of putative risk factors, such as exposure to mutagens,⁵⁻¹² inbreeding¹³ and environment.^{7,14}

CHD patients have been referred to the Cardiovascular Department at the King Faisal Specialist Hospital and Research Centre (KFSHRC) since its inception in 1977. An Internet-based CHD registry (CHDR) was established in 1998 for both clinical and research purposes.¹⁵ By linking the CHDR data to a geographical information system (GIS) currently used by the Saudi Arabian Ministry of Defense and Aviation (MoDA), we have been able to explore the geographical distributions of CHDR patients. A GIS is a software package dedicated to the storage and display of geographic information. This paper focuses on the development of the system and the initial results. Since there appear to have been no prior GIS publications related to health care or epidemiology in Saudi Arabia, it may be worth mentioning that this system can be adapted to study the geographical distribution of other diseases.

Methods

Definition of CHD. The CHDR routinely codes patients according to both ICD-9-CM (745.0 to 747.9, including Wolf Parkinson White syndrome, 426.7, and supraventricular tachycardia (SVT), 427.89)¹⁶ and the newly established European Pediatric Cardiac Code.¹⁷ No effort is made to distinguish congenital from acquired SVT. Patients with isolated patent ductus arteriosus (PDA) diagnosed at less than 90 days of age are excluded, and cases of mitral valve prolapse are not registered.¹⁸⁻¹⁹ All cases were confirmed by echocardiography, cardiac catheterization or cardiac surgery (autopsy within Saudi Arabia is performed only in exceptional circumstances). The CHD cases were analyzed as a group to avoid errors that might result from the small number of cases for some specific diagnoses.

Map data. The GIS themes (i.e., sets of related geographical features) presented here were obtained in their entirety from the Saudi Arabian Ministry of Defense and Aviation, with the exception of the cities of CHDR patients; these were supplemented by information from the "Digital Chart of the World," an Environmental Systems Research Institute, Inc. (ESRI) product that was freely downloaded across the Internet. The MoDA cities theme initially comprised 994 entries, but after eliminating locations that were not habitations (i.e., had no population), and multiple entries for identical x,y coordinates, 970 unique city names remained.

Data analysis. GIS analysis was carried out using the ArcView software package (v3.2a, ESRI Systems Inc.). Other data-analysis made use of the JMP statistics package (v3.2.5). The scatterplots, line-plots and histograms were produced by Origin v5.1 (Microcal).

Independence of measurement. Families with more than one case of CHD in the registry were located using a family membership identifier (assigned to each case upon registration, and based upon information from the parents), in conjunction with a retrospective computer search of the entire registry (using family name, grandfather's name, father's name, telephone number, province and cities of residence and origin). When multiple children with CHD from the same family were detected, only the first-born child was retained.

Demographic data. The city and province of current residence (i.e., the residence of the father at the time of registration) were obtained from a parent of the child during a face-to-face interview. Within the Kingdom, this is expected to closely reflect the mother's residence and will be largely unaffected by the location at which the woman actually delivers. Since most cases were reg-

istered very early in life and the pool of migrant indigenous workers has traditionally been small, this should be a good indicator of where the child was gestated. The term "city" applies here to a specific geographical location, as opposed to the more general location indicated by the administrative province; this may refer to an actual city, or a town, or even a small village. Estimates of the Saudi populations of each city were taken from the 1413 Hejra (1992-1993 Gregorian) census.²⁰

Results

Description of CHD Cases. The patient population comprised all 6649 patients registered in the KFSHRC Congenital Heart Disease Registry (CHDR) between 1 January 1998 and 1 November 2002. Five hundred and twenty-two non-Saudi cases, 51 subsequent registrations from the same families, 84 cases resident outside the Kingdom of Saudi Arabia (KSA) at the time of their interview, and 127 cases which were never interviewed, were all excluded, leaving 5865 cases for further study. The province-of-residence was known for all 5865 cases, and the city-of-residence was available for 5764, of which only 5209 could be successfully linked to a specific geographical location (Table 1).

Construction of the GIS Map (Provinces). In both the CHDR and the MoDA databases, province names were coded as English transliterations of the original Arabic names. Although there was some variation in spelling, each province was easily identified because there were only 14 different entities. The names of the provinces in the MoDA database were modified to correspond to those used in the CHDR. During this process it was observed that the CHDR data contained one additional province, Qurayyat, which the MoDA database had included as part of the Jawf province. This situation arose because of changes in the administrative boundaries during the last 10 years. The CHDR designation was changed from Qurayyat to Jawf.

Construction of the GIS Map (Cities). This CHDR dataset contained 186 unique names for city-of-residence and 162 for city-of-origin, which together comprised 202 unique city-names. The original intention was to obtain the geographical coordinates of these cities by automatically screening each name against the MoDA database of 970 cities with their associated longitude and latitude. As with the province names, the city names had been coded (in both databases) as English transliterations of the original Arabic. However, since there were so many more city-names than province-names, the different transliterations meant that it was not possible to automatically match each of the CHDR

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Table 1. Distribution of cases in the KFSHRC Congenital Heart Disease Registry among the different provinces.

Province Name	Saudi population	Number of cases		
		Total cases	Cities with known geographical location	Cities with unknown geographical location
Asir	1 149 618	349	296‡	53‡
Al Baha	289 890	140	128	12
Eastern Province	1 898 462	1 469	1 464	5
Hail	346 180	41	38	3
Jawf	224 040	138	138	0
Jizan	734 078	202	193	9
Medina	836 764	369	363	6
Makkah	2 780 458	1 054	1 051	3
Najran	242 066	161	157	4
Northern Province	178 389	79	79	0
Qasim	611 462	241	156¶	85¶
Qurayyat†	n/a	82	79	3
Riyadh	2 613 228	1 300	1 286	14
Tabuk	401 256	240	233	7
Total	12 305 891	5 865	5 209	66

* Population statistics taken from the last published census, 1413 Hejra (1992-1993 Gregorian).

† For analysis, Qurayyat province has been included in the statistics for Jawf province.

‡ 44 cities of residence for Asir had invalid codes.

¶ 59 cities of residence for Qasim had invalid codes.

cities to its corresponding MoDA entry. After a simultaneous match-merge operation using province and city names, 82 of the 202 cities (approximately 40%) remained unidentified. Sixty-nine of these were subsequently identified by closely inspecting the transliterations of each CHDR city-name and (where possible) manually identifying the corresponding name in the MoDA database. Five more cities were identified using an alternative city database and the names of the eight remaining cities whose geographical coordinates could not be identified were set to missing (19 CHDR cases, 11 cities-of-residence and 16 cities-of-origin). Two pairs of cities were discovered to have identical longitude and latitude in the MoDA database (Ad Dammam/Dammam and Ballahmar/Ballasmar); Dammam and Ballahmar were therefore deleted from the list of city names, and their six corresponding entries in the CHDR dataset were also changed. Two hundred city names remained in the final CHDR dataset.

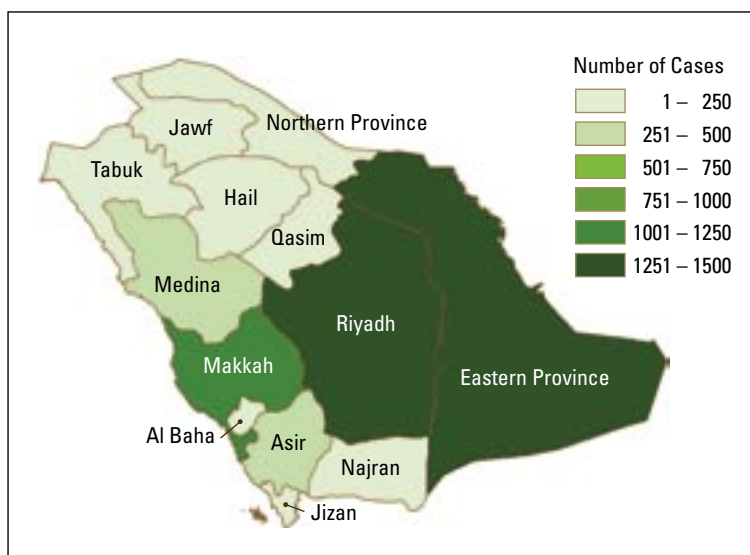


Figure 1. Geographical distribution of KFSH&RC congenital heart defect cases among the provinces of Saudi Arabia (1998-2002).

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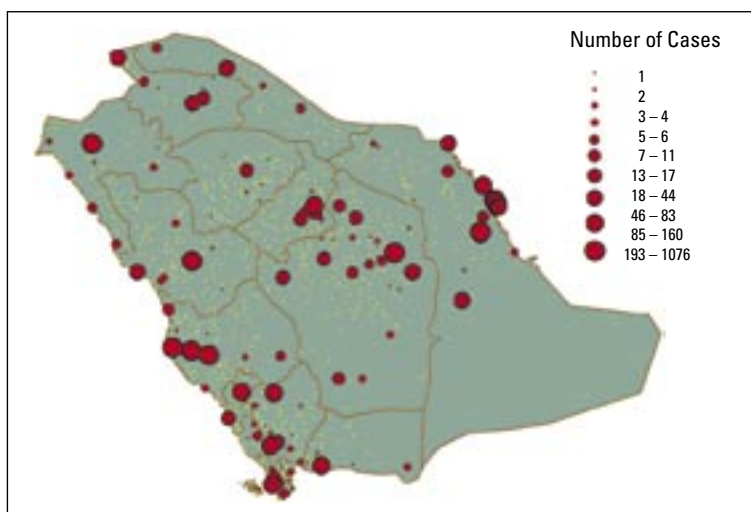


Figure 2. Distribution of KFSH&RC congenital heart defect cases among the cities of Saudi Arabia. (Cities classified into ten quantiles. Cities with no cases shown as small yellow dots.)

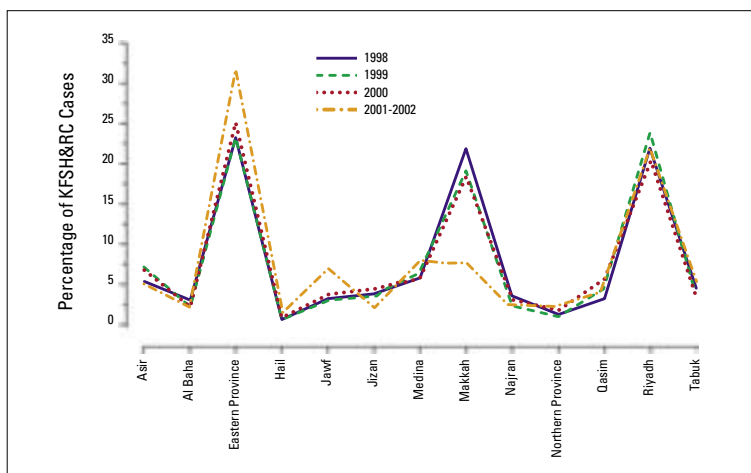


Figure 3. Percentage distribution of KFSH&RC congenital heart defect cases registered each year in the provinces of Saudi Arabia. (Cases for 2001-2002 are incomplete)

Table 2. Distribution of registered congenital heart disease cases by year.

Year	Number of registrations
1998	2053
1999	1707
2000	1125
2001	732*
2002	248*

* Not all cases registered in 2001 and 2002 had been processed when the dataset was abstracted.

Geographical distribution of CHDR cases. The geographical distribution of CHD (Figure 1) reflected the regional population density. The largest number of cases were from the Riyadh, Eastern and Makkah provinces, forming an east-west “axis” across the centre of the country. However, due to the large size and inhomogeneity of the Saudi provinces, the population densities underlying these maps were not uniformly distributed within each province, thereby creating a misleading impression of the true geographical distribution. In a thematic map based on cities-of-residence (Figure 2), the “axis” is less obvious and is more clearly focused only around the 3 largest cities—Dammam (east coast), Riyadh (central) and Jeddah (west coast). What was not so evident from the map of the provinces is that there is also clustering of cases.

Registry stability and coverage. The CHDR has been functional since 1998. Due to the existence of a sizable and well-established outpatient population, more than 2000 new cases were registered during its first year of operation (Table 2). The number of new cases registered each year has subsequently declined to approximately 1000. However a time-series of thematic maps (not shown) suggested that the geographical distribution has remained very similar on an annual basis. The annual change in the percentage of new registrations appeared similar across the provinces (Figure 3), and this was confirmed by the corresponding thematic map for cities (Figure 4), between the years 1998 and 2000. A comparison between the number of CHD cases per province, and the population of each province at the 1413H census (Figure 5) lends support to the notion of representative CHDR coverage, except for the Eastern Province which has a significantly higher number of cases.

The CHD burden. A geographical analysis based solely on numbers of cases can reach only limited conclusions. To obtain a more accurate picture of the disease distribution, the sizes of the underlying city populations need to be considered. A map of prevalence estimates would be ideal, but currently there is only limited available information for birthrates at the level of detail required. An indication of the underlying CHD “burden” (CHDB) can be estimated by dividing the number of cases for each province or city by its population (expressed per 100 000). Although this is not a true prevalence measure, it does provide a convenient way to normalize the results. Because this estimate is based on 1413H census data, those CHD cases born outside a “window” of ± 5 years around

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1413H were excluded from this analysis, resulting in a 10-year subset of 3151 CHDR patients (distributed across 139 cities) whose birthdays occurred between 1988 and 1997 inclusive. A further 109 cases were eliminated because their city-of-residence was either missing (11 cities, 45 cases) or had an unknown location (3 cities, 64 cases), leaving 3042 cases distributed across 125 cities for further study. The Saudi population could not be definitively established for a further 19 cities (15%) leading to missing CHDB estimates. The resulting distribution of CHDB (Figure 6) was positively skewed and contained only one “outlier” -Al Baha (in the Al Baha province) with a CHDB estimate of 748 per 100 000 (70 cases from a population of 9364). This city lies in the southwest, and an inspection of the geographical distribution of CHDB (Figure 7) showed that the southwest provinces as a whole appeared to be the region that had the largest number of cities with a substantial CHD burden, although several cities in the Eastern Province were also affected.

Discussion

As far as we are aware, this is the first publication of a detailed GIS suitable for epidemiological research in Saudi Arabia. Paradoxically, the major obstacle in applying GIS to local health care issues is not a lack of accurate geographical data (indeed a number of GIS maps of the Kingdom can be downloaded free from the Internet²¹), but rather the problem of establishing the appropriate linkages between the map and the project databases. This is partly a language problem: data in key fields (in either database) can be in English, so that success in merging such data depends upon how consistently these have been transliterated. Even in Arabic, the names of the smaller towns or villages can be ambiguous because the same city may be known by different names. There is also a problem in obtaining accurate demographic data, because the population structure has changed so quickly since the last published census (10 years ago) that simple interpolation is inappropriate, and there is no guarantee that data from different ministries can be correlated (e.g., different administrative boundaries may be used).

This is also the first attempt to portray the geographical distribution of CHD in KSA, and our preliminary maps have revealed several important features. Although the CHDR population distribution reflects the central axis of population, cities in

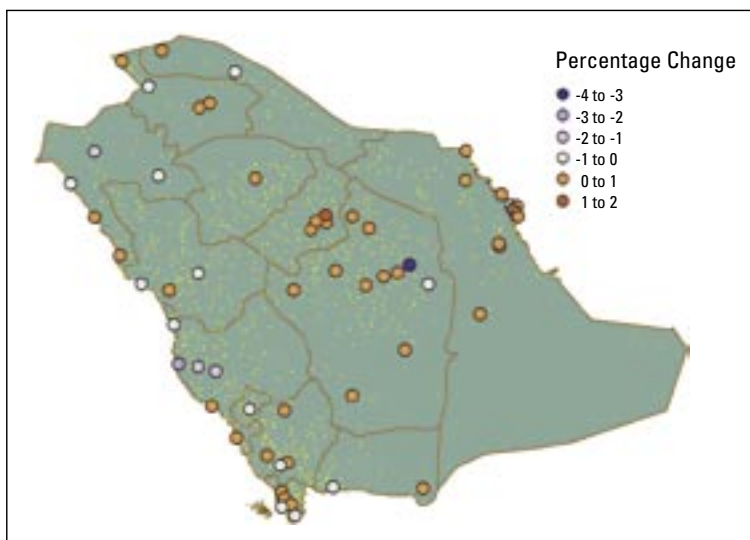


Figure 4. Change in percentage distribution of KFSH&RC congenital heart defect cases among the cities of Saudi Arabia, registered 1998-2000. (Small yellow dots represent cities with no cases.)

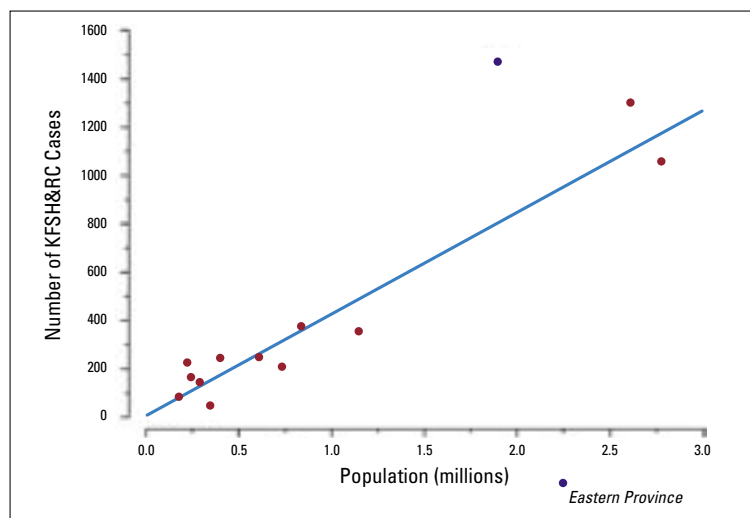


Figure 5. Number of KFSH&RC congenital heart defect cases per province vs. the population of each province in the 1413H census.

the southwestern provinces (Jizan, Asir, Najran and Al Baha) exhibit high disease burdens. Indeed, Al Baha city appears to have the largest CHD burden of any city in the Kingdom, although since the city has the same name as the province, the possibility of some data-capture errors cannot be ruled out. The CHD problem in the southwest may reflect the unique physical terrain of that region, which is more mountainous than other parts of the Kingdom. Alternatively it may be associated with cultural

factors that have been imported from its southern neighbor, Yemen. Traditionally, the Kingdom's border has been more permeable here than elsewhere in the Kingdom.

The value of these results depends strongly on the extent to which the CHDR data represents the entire Kingdom. This is admittedly difficult to gauge since it depends largely upon the referral pattern. However, for a rare disease such as CHD we would argue that the existence of a small number of referral

hospitals should guarantee a nationwide dimension. Apart from KFSHRC, there are currently only two other hospitals in KSA that treat CHD patients. Our current estimate is that KFSHRC alone captures approximately 50% of the total CHD burden. Since the cardiovascular department at KFSHRC has been treating CHD cases for more than 15 years before the registry began, it is likely that the catchment area would have become stabilized, and our results appear to confirm this.

There are several minor factors that may have introduced some bias. It is possible that some cases have not been correctly identified, especially in the provinces. This, and the recent growth of the hospital's "Outreach" program might have led to a degree of inhomogeneity. There were also a small number of cases that were excluded because their geographical location could not be determined. However, the cities for which we could obtain no reliable population statistics represent 15% of the final dataset. This constitutes the most significant source of error in this study.

We have also shown that displaying spatial distributions in KSA using maps based on provinces alone is inadequate in faithfully representing disease distributions in a geographical area as inhomogeneous and sparsely populated as Saudi Arabia. Maps based on cities convey a more accurate impression. Furthermore, by constructing maps of the disease burden only for those cities that actually contain cases, not only do we improve the accuracy of the spatial distribution, but we also minimize any residual bias due to referral patterns by not including populations in the denominator that are not demonstrably included in the catchment area of the registry.

In conclusion, we believe that we have produced a GIS system that is sufficiently accurate to tackle the problem at hand (the spatial distribution of CHD) which can be extended to other disease registries or national studies by including information as it is made available. We hope that through time, this can evolve into a comprehensive epidemiological GIS for the Kingdom of Saudi Arabia.

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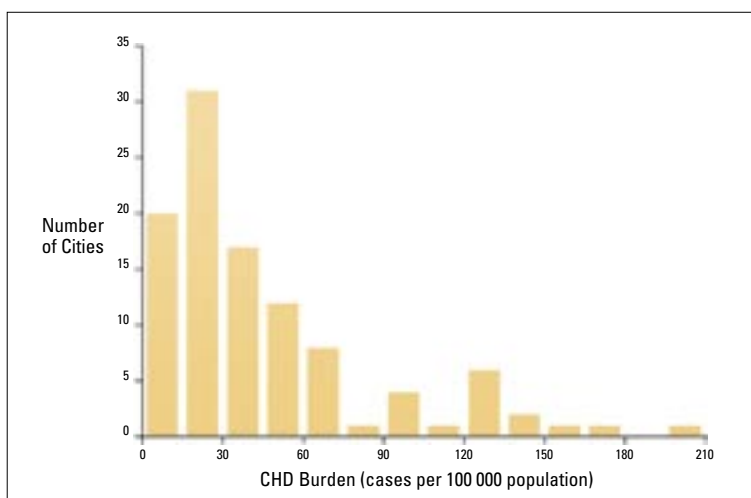


Figure 6. Congenital heart defect burden of KFSH&RC CHD cases among the cities-of-residence for cases with birth-years between 1988 and 1997 (inclusive). (Al-Baha is regarded as an outlier and therefore not included.)

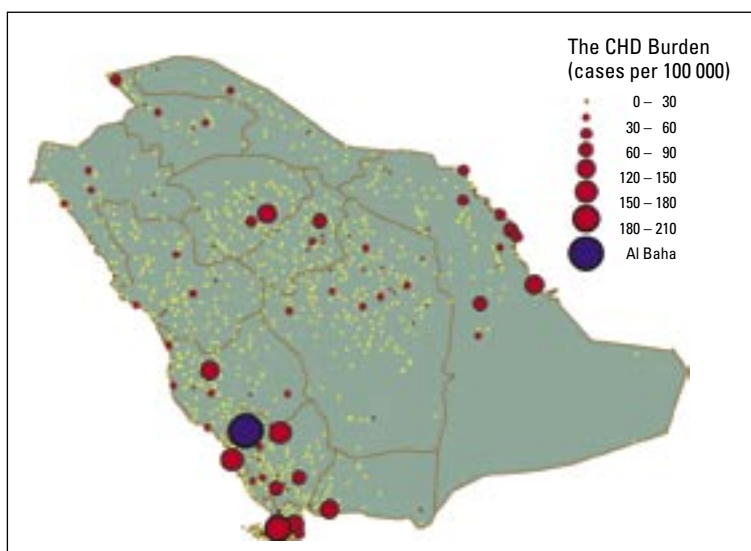


Figure 7. Congenital heart defect burden for KFSH&RC CHD cases with birth-years between 1988 and 1997 (inclusive), distributed among cities-of-residence. (Al-Baha shown in blue because it has a disproportionately high value. Cities with no cases shown as small yellow dots.)

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