National Congenital Abnormalities Registry in UAE (1999-2004)

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Brief findings

In the United Arab Emirates the rate of infant mortality decreased from 11.37 per 1000 live births in 1990 to 7.8 per 1000 live births in 2003.

However , the proportion of infant mortality due to congenital abnormalities (CAs) showed an increasing trend ; this proportion was 86.1% in 2002 which was increased from the previous 30% in the late 1980s.

Justification

The high proportion of infant mortality caused by CAs (86.1%) means that ***prevention*** of CAs and establishment of national congenital abnormalities registry was a high priority in the United Arab Emirates.

CAs represent a special category of disorders characterized by their earliest onset and limited chance for complete recovery. ***Prevention*** , which is the only optimal solution, is based on real knowledge of baseline prevalence of different CA entities, causes of CAs and possible risk factors.

Objectives of the registry

a. To determine the baseline birth prevalence of different congenital abnormality entities .

b. To highlight the topic for the medical community in order to improve the quality of diagnosis and recording.

c. To establish a priority list of preventive efforts .

d. To use the surveillance function of the registry .

Protocol of Registry

National Congenital Abnormalities Registry (NCAR) is ***population based*** covering all births in UAE and was established in all medical districts in January 1999

Congenital abnormalities are *structural defects* of fetal development with necessity of medical treatment. So minor anomalies such as hydrocele and preauricular tag are excluded.

Congenital Abnormalities Registry (CAR):
CAR is a *system* of ongoing and permanent registration for the collection, storage and analysis of personal, demographic and medical data on affected neonates and infants.

Cont.

Protocol of Registry

•Study period:

- From birth till the age of 1 year (stillbirth, live birth) .
- From birth till the age of 12 years for hereditary disorders .

Source of information: In the nine medical districts of UAE

·All maternity obstetric units (99% of deliveries in hospital) .
·Pediatric clinics (neonatal, general, surgical) .
·MCH & PHC centers . . Genetic laboratory .
·Hematology Clinics . . Hearing and Deafness Clinics .

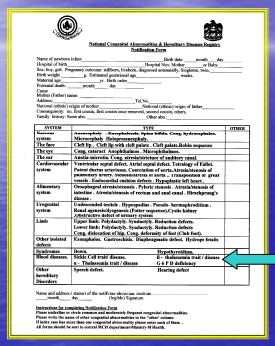
Cont.

Protocol of Registry

Method of Notification: Printed notification form .

Unit of recording: Index cases with isolated and multiple congenital abnotmalities .

Classification of cases: According to the codes of the international classification of Diseases (ICD) 10th version (Q00.0-Q99.9) with slight modification in multiple CAs .



NEW

Notification Form

The new notification form contains also the common hereditary disorders selected from the hereditary registry.

Plan of Action

There are many steps and studies have been taken :

1- National Committee: for hereditary diseases in 1994.

2-WHO (EMRO) continously provided technical support through STC.

Dr.Andrew Czeizel: helped in *assessing* the situation in the UAE, *in preparing the staff* for the registry, developing the *registration form* and lastly *evaluated* the start of the national congenital abnormalities registry.

Cont.

3- Studies which have been done:

- *Retrospective study* in Al Mafraq Hospital between 1992 and 1994 to evaluate the problem *(published in Congenital Anomalies J. , 1999).*
- *Field testing* to check the practical use of the notification form in Al Mafraq and Al Cornish hospitals .
- ** Pilot study* of National congenital anomalies registry in 3 medical districts (Abu Dhabi, Al Ain and Western Region) in 1998 to finalize the notification form and to determine the baseline birth prevalence of different CAs *(published in Teratology J., 2000).*

4- National Congenital Abnormalities Registry :

started in all medical districts since January 1999 *(published in Eastern Mediterranean Health Journal , 2005).*

Results

A total number of 346,728 total births (LB+SB) were included since the implementation of the NCAR (from January 1999 until December 2004) .

	Live births	Stillbirths	Total births
No. of Births	344,167	2561	346,728
No. of CA	4793	125	4918
Prev. per 1000	13.9	48.8	14.2

Table (1) Prevalence of reported congenital abnormality (CA) according to the pregnancy outcome , 1999-2004 .

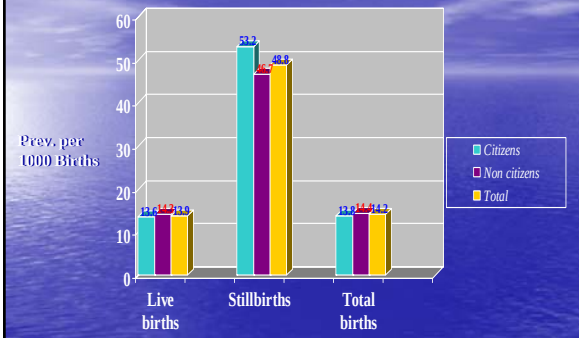


Fig. (1) Prevalence of reported congenital abnormality (CA) according to the nationality (1999-2004) .

Type of CA	Pregnancy outcome					
	Live births		Stillbirths		Total births	
	CAs, No.	No./ 1000	CAs, No.	No./ 1000	CAs, No.	No./ 1000
A-Isolated CAs	1316	9.1	25	32.5	1343	9.3
1) Neural-tube defects	140	0.9	13	16.9	153	1.1
2) Other CAs of Nervous System	72	0.5	4	5.2	76	0.5
3) CAs of Eye	14	0.1	0	0.0	14	0.1
4) CAs of Ear, Face and Neck	23	0.2	0	0.0	23	0.2
5) CAs of CVS	378	2.7	4	5.2	382	2.7
6) CAs of Respiratory System	9	0.1	0	0.0	9	0.1
7) Orofacial Cleft	86	0.6	4	5.2	90	0.6
8) CAs of Digestive System	63	0.4	0	0.0	63	0.4
9) CAs of Geneital Organs	180	1.2	0	0.0	180	1.2
10) CAs of Urinary System	90	0.6	0	0.0	90	0.6
11) Certain CAs of Muscloskeletal	95	0.7	0	0.0	95	0.7
12) Other CAs of limbs	86	0.6	0	0.0	86	0.6
13) Other CAs of Muscloskeletal	63	0.4	0	0.0	63	0.4
14) CAs of Integument	9	0.1	0	0.0	9	0.1
B-Multiple CAs	641	4.5	16	20.8	657	4.5
1) Gene CAs	131	0.9	0	0.0	131	0.9
2) Chromosomal CAs	252	1.7	0	0.0	252	1.7
3) Other Multiple CAs	14	0.1	4	5.2	18	0.1
4) Unspecified multiple CAs	244	1.7	12	15.6	256	1.8
Total	1959	13.6	41	53.2	2000	13.8

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Table (2) prevalence of different categories of congenital abnormality (CA) according to the pregnancy outcome for United Arab Emirates citizens, 1999-2004 .

Type of CA	CAs, No.	No./ 1000
A-Isolated CAs	519	13.8
Neural-tube defects	54	1.5
Other CAs of Nervous System	36	0.9
CAs of Eye	10	0.3
CAs of Ear, Face and Neck	7	0.2
CAs of CVS	167	4.6
CAs of Respiratory System	2	0.1
Orofacial Cleft	94	2.5
CAs of Digestive System	19	0.5
CAs of Geneital Organs	44	1.2
CAs of Urinary System	3	0.1
Certain CAs of Muscloskeletal	36	0.9
Other CAs of limbs	24	0.7
Other CAs of Muscloskeletal	19	0.5
CAs of Integument	4	0.1
B-Multiple CAs	104	3.3
Gene CAs	20	0.6
Chromosomal CAs	61	1.7
Other Multiple CAs	9	0.3
Unspecified multiple CAs	14	0.4
Total	623	17.1

Table (3) prevalence of different categories of congenital abnormality (CA) per 1000 total births for United Arab Emirates citizens in Abu Dhabi medical district, 1999-2004 .

Year	/1000 Total births
1999-2003 (NCAR)	1.48
1992-1999 (Samson, etal)	1.13
1996-2000 (Sedaghatian, etal)	1.77

Table (4) Comparisons with other Neural Tube Defects studies done for United Arab Emirates citizens in Abu Dhabi medical district.

Risk Factors

- Maternal variables.
- Infant variables.

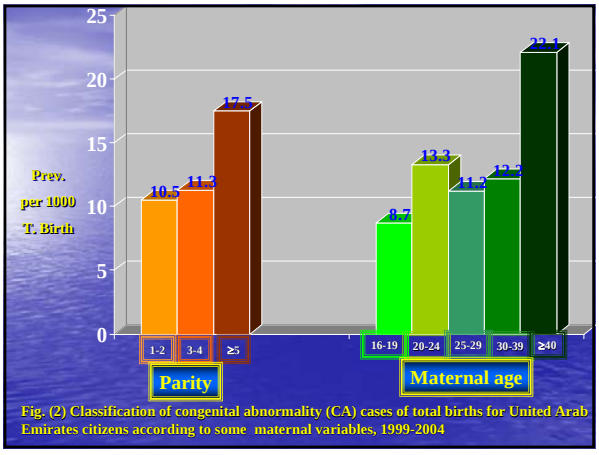

Fig. (2) Classification of congenital abnormality (CA) cases of total births for United Arab Emirates citizens according to some maternal variables, 1999-2004

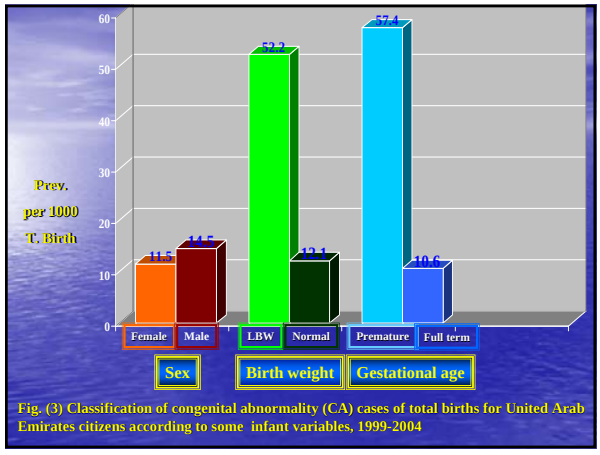

Fig. (3) Classification of congenital abnormality (CA) cases of total births for United Arab Emirates citizens according to some infant variables, 1999-2004

Prevention

PREVENTIVE GENETIC COUNSELING PROGRAMS :

1 - Neonatal Screening Program :
PKU , Cong Hypothyroidism , Sickle Cell Diseases and Congenital adrenal hyperplasia .

2 - Congenital Abnormalities & Hereditary Diseases Registry .

3 - Genetic Clinic

4- Genetic Laboratory

5 - Premarital Counseling Program

Risk Factors
Maternal age
Pregnancy care
Rubella
Self medication
Smoking
Folic acid

Obstacles (Constrains)

1- Under ascertainment of malformation cases, because the doctors in some hospitals are not interested in registration.

2-Registry includes only live born children and rarely stillborn.

3- Inadequate Filling of the form : - Missing information e.g. consanguinity , hospital of birth, etiology and in some occasions, babies over one year of age are recorded.

Recommendations To Improve Program Effectiveness

- Increase ascertainment by using multiple-sources case finding e.g. birth certificates, hospital activity analysis register, perinatal death certificates and genetic clinics .

•Enhance the knowledge and skills of the health professionals especially those in medical districts where underreporting is an obvious concern by continuous short courses, practical workshops and lectures .

•Also, it is important to be connected to one of super national organization of CAs registry programs as International Clearinghouse for Birth Defects Surveillance and Research (ICBDSR) .

The Clearinghouse is a voluntary nonprofit organization affiliated with the World Health Organization (WHO).

The mission of the International Clearinghouse for Birth Defects Surveillance and Research is to bring together birth defect programs from around the world with the aim of conducting worldwide surveillance and research to prevent birth defects and to ameliorate their consequences.

FUNCTIONS

A. Operate an international program for regular exchange among its members of information on birth defects in populations covered by the member's surveillance and research programs.

B. Cooperate in investigations and research into changes in the occurrence of birth defects.

C. Conduct joint epidemiological studies of the causes of birth defects.

D. The Clearinghouse will promote standards and definitions for conducting surveillance of various types of birth defects.

E. Provide effective training in the surveillance and research of birth defects.

F. Be an advocate for the surveillance, research and prevention of birth defects.

G. Conduct assessments of preventive and therapeutic interventions for birth defects.


Thank you