

Original Article

Designing an Electronic Registry for Patients with Beta Thalassemia Major for Mazandaran Province, IR Iran 2016

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Abstract

Background: Beta thalassemia major is the most prevalent genetic disease in Mazandaran province. The national prevention program is successful, however our patients grow older and their managements get more complicated. Paper based clinical notes are being used and there are some electronic data banks mainly for financial matters. This registry (ETR Mazandaran) is designed to be a reliable data bank for epidemiologic and clinical data for beta thalassemia major patients living in the province. This registry will be a source for health system decision makers.

Methods: Thalassemia Research Center (TRC) with collaboration of informatics Technology Department of Mazandaran University of Medical Sciences (MUMS) designed an Electronic Thalassemia Registry (ETR, [Http://thr.mazums.ac.ir](http://thr.mazums.ac.ir)) in Persian language, in 2015-2016. There are 16 dedicated clinics and wards under MUMS in the province taking care of about 2700 patients. In each thalassemia clinic a nurse has been elected and educated. Questionnaires were designed by an experienced clinician with collaboration of a software developer. Each patient was registered by the national identification number and by his (her) name. Epidemiologic, clinical data, complications and medication have been registered. Confidentiality has been considered and no reports include patient's name. All research proposals were ethically evaluated in TRC research committee and university ethical committee if involves intervention in patients management. A biologic bank for blood or DNA samples has been anticipated.

Results: A Web-based Thalassemia Registry system was designed to serve as an electronic medical file for patients retrieving data from medical files by trained registrars. Data was audited and cleaned before it was

archived in the electronic filing system. The registry is consisted of epidemiologic data, clinical history and important physical examination points, laboratory results, a list of classic complications, list of medication. Each record should be up dated at least one time in each season. There are also a separate form for special checkup such as echocardiogram, MRI T2* and bone mineral densitometry which should be updated annually for patients 16 years old or older. There is a form for migration to other provinces and death. This registry is designed to run for at least 10 years. Thalassemia registry is easily adaptable for usage in other provinces.

Conclusion: Electronic Thalassemia Registry is an online data bank for patients, physicians and policy makers for better research and development in the field.

Key words: Beta Thalassemia Major, Electronic Registry, Mazandaran, IR Iran

Background

Beta thalassemia major is the most prevalent genetic disease in Mazandaran province, as well as the country. The national program for prevention of new cases is successful in the province, however, due to better clinical management our patients grow older and their managements get more multi-disciplinary and complicated (Abolghasemi et al., 2007, Khorasani et al., 2008, Hashemieh et al., 2015).

There is a national guideline for management of patients with beta thalassemia major in Islamic Republic of Iran which was distributed in 2006 (Azarkeyvan A, 2006). Paper based clinical notes are being used and is the main source for our researches (Kosaryan et al., 2007, Karami et al., 2010). However, in some centers medical notes are incomplete and inaccurate.

There are some electronic data banks mainly for financial matters. At least 2700 patients live in the province and are being managed in 17 hospital based clinics. Some 80% (2160 patients) of patients are under Mazandaran University of Medical Sciences (MUMS), however, 20 % (540 patients) are being cared in one hospital under Babol University of Medical Sciences. This registry (ETR Mazandaran) is designed to have a reliable data bank for epidemiologic and clinical researches for the cohort of patients with beta thalassemia major living in the province. This data is needed for research purposes as well as routine health policy making. This registry will be a reliable source for patients, doctors and also health system decision makers.

Methods

Thalassemia Research Center with collaboration of Technology Department of the university designed an Electronic Thalassemia Registry (ETR, [Http://thr.mazums.ac.ir](http://thr.mazums.ac.ir)) in 2015-2016. The registry was developed in agreement of the national guideline for management of patients

with beta thalassemia major (Azarkeyvan A, 2006).

Governing body: A responsible committee consisted of the head of TRC with two other members including an epidemiologist are responsible for design as well as financial matters. Acceptance of research projects proposed by persons outside of the center is also subject to this committee.

Geographic definition: In this phase the registry is for patients who are living in Mazandaran province. All cities are set as default places, also, the name of hospitals which has thalassemia clinics are available.

Hospital settings: Meanwhile, there are 14 hospital based dedicated thalassemia wards and clinics under Mazandaran University of Medical Sciences in the province.

Patient eligibility: All patients with the diagnosis of beta thalassemia major by the responsible physician of thalassemia clinics, according to CBC and hemoglobin electrophoresis are eligible for registration.

Data collection forms: Data collection forms were designed in Persian, by an experienced clinician with collaboration of a software developer during consulting sessions. In each thalassemia clinic a nurse has been elected and educated for data entry.

Confidentiality: Different level of access to data has been defined. Patients could access only to their data, however, they could not edit the data. In each hospital a dedicated operator have access to the data of all patients in the particular center and is able to change data. Central admin has access to all data and is able to create statistical reports.

Links: Software has links to different related bodies including site of MAZUM, Mazandaran Thalassemia Society (Charity).

Biologic bank: A biologic bank for blood or DNA samples has been anticipated in molecular laboratory of TRC located in Sari.

Quality control of filled forms: Admin is able to randomly select 5% of registered patients to data control. Directions for filling the forms could be found in a dedicated part. There are also short directions in each part upon pause of the cursor. Data base acceptability and technical problems: Data base is accessible only for statistical reports by headquarter of system. Language of program is English. Region of server is in Sari. Security issues and technical problems will be covered by the designing body. Descriptive statistics, as well as, pie and column diagrams could be created for all variables for each center according to patient's gender. This report can be saved, mailed or printed.

Guidelines and answers for questions: There are 3 levels of guidance for nurses responsible for each ward and clinic. The first line is a small description while the cursor remains still on most variables. Also there is a separate information part in the dashboard. There is the possibility to ask questions and get the answer publicly or private for all nurses by an intra-mail system. We also performed a teaching session for all participating nurses. We also informed all physicians in a scientific meeting by a lecture.

Variables: Demographic data consisted of; name, gender, fathers name, place and date of birth, identification number, address and telephone number, education and occupation, marital status, type of medical insurance, consanguinity of the parents, age of first transfusion, blood type. As the national program for prevention of thalassemia started in 1997, births after this date should be asked about the reason for the failure of preventive acts by one of three options; PND (prenatal diagnosis) was not performed, was not accurate, abortion was denied by any reason.

Data for physical examination page is consisted of the weight (Kg), hepatomegaly (yes/no), growth problem (yes/no), Facial disfigurement (yes/no), history of splenectomy (yes/no), if spleen was in place, if there is splenomegaly (yes/no) is going to be registered.

Page for thalassemia complications is consisted of data about being affected by, bile stone,

diabetes mellitus, cardiomyopathy, hypothyroidism, osteoporosis, hypoparathyroidism, and fertility disorder, all as yes or no options.

Page for seasonal laboratory results is consisted of the mean pre transfusion hemoglobin levels, FBS, Ferritin, Ca, AST, ALT, P and blood urea.

Annual laboratory results are consisted of; TSH, FBS, blood glucose 2 hours after meal as gram / deciliter. Also the result of glucose tolerance test (Not indicated/ not done/ normal/ abnormal). HCV, HIV antibody, HBS antigen and antibody titer And also serum zinc.

Vitamin D level, bone mineral density, MRI T2* of heart and liver, echocardiogram, all by three options; yes/no/not indicated.

Results of MRI T2* is going to be registered as normal, mild, moderate and severe categories. Result of bone mineral density is going to be registered as Z scores of total femoral neck and total lumbar vertebra.

Treatment and medication page is consisted of if the patient is under blood transfusion. The type of blood that the patient was using (leukoreduced/ washed). Adverse reaction to the blood and presence of antibodies (as yes/no options).

An alphabetical list of popular medications has been anticipated that could be ticked. There is possibility to add more medications to the list.

There is a page for migration to other provinces. Also the death (with the date) is going to be registered. There is an option to choose between deaths related to thalassemia or unrelated (accidents).

Results

Registry has 7 different pages for epidemiologic data, important physical exams, complications, seasonal laboratory data, annual laboratory data, blood and medication information list and migration (death event form) (Figure. 1-7).

Stared variables are mandatory to be filled in order to pass the page. Software has alarm system for them. For all quantitative variables a range of possible digits has been anticipated and an alarm system will be activated upon out ranged measures.

Demographic Information	
Date of entry	[ShowPresentTime]
File Number	<input type="text"/>
National Code	<input type="text"/>
Name	<input type="text"/>
Family	<input type="text"/>
Gender	Mail <input type="radio"/> Femail <input type="radio"/>
Birth Date	<input type="text"/> <input type="text"/> <input type="text"/>
Province of birth	<input type="text"/>
City of birth	<input type="text"/>
Province Location	<input type="text"/>

Figure 1: The page for demographic in in electronic registry for patients with beta thalassemia major for Mazandaran province, IR Iran 2016.

physical examinations

Year of PhyExamination	<input type="text"/>
Season	<input type="text"/>
Weight	<input type="text"/> Kg
Hepatomegaly	<input type="text"/>
Facial disfigurment	<input type="text"/>
Growth failure	<input type="text"/>
Splenectomy	<input type="text"/>

Figure 2: The page for physical examinations in electronic registry for patients with beta thalassemia major for Mazandaran province, IR Iran 2016.

Important Thalassemia Complications

Year of Lab Result	<input type="text"/>
Season	<input type="text"/>
Bil. stone	<input type="text"/>
Diabetes	<input type="text"/>
Cardiomyopathy	<input type="text"/>
Hypothyrodism	<input type="text"/>
Osteoprosis	<input type="text"/>
Hypoparathyrodism	<input type="text"/>
Fertility disorder	<input type="text"/>

Figure 3: The page for important thalassemia complications in electronic registry for patients with beta thalassemia major for Mazandaran province, IR Iran 2016.

Seasonal Laboratory Data

Year of Lab Result	<input type="text"/>	
Season	<input type="text"/>	
Mean of pre-transfusion Hb	<input type="text"/>	gr/dl
FBS	<input type="text"/>	mg/ml
Ferritin	<input type="text"/>	ng/ml
Ca	<input type="text"/>	mg/dl
AST	<input type="text"/>	u/l
ALT	<input type="text"/>	u/l
P	<input type="text"/>	mg/dl
Urea	<input type="text"/>	mg/dl
Uric Acid	<input type="text"/>	mg/dL

Figure 4: The page for seasonal laboratory data in electronic registry for patients with beta thalassemia major for Mazandaran province, IR Iran 2016.

Annual Laboratory Data

Year of Lab Result	<input type="text"/>	
TSH	<input type="text"/>	mIU/dL
F.B.S	<input type="text"/>	mg/dl
BS(2H)	<input type="text"/>	mg/dl
OGTT	<input type="text"/>	
HCV antibody	<input type="text"/>	
HIV antibody	<input type="text"/>	
HBS antigen	<input type="text"/>	
HBS antibody	<input type="text"/>	
Zinc	<input type="text"/>	mg/dl
Vit. D	<input type="text"/>	ng/mL
PTH	<input type="text"/>	mg/ml
MRI T2*	<input type="text"/>	

Figure 5: The page for annual laboratory data in electronic registry for patients with beta thalassemia major for Mazandaran province, IR Iran 2016.

List of Medicine:

- ASA
- Calcium
- Calcitriol
- Digoxin
- Estrogen
- Folic acid
- Flosemide
- Insulin
- Levothyroxin
- Oral hypoglysemic
- Penicilin
- Progesterone
- Testosterone
- Vit.D
- HydroxyUrea
- Allupotinol
- Propranolol
- Syndenafil

Med. for osteoporosis

Iron chelator

DFO

DFX

DEF

Figure 6: The page for medications in electronic registry for patients with beta thalassemia major for Mazandaran province, IR Iran 2016.

DFO: deferoxamine, DFX: deferasirox, DEF: deferiprone

Migration and Death

The image shows a web interface for data entry. At the top, there is a horizontal line. Below it, on the left, is a dropdown menu with a downward arrow. To its right is the label 'Patient State'. Below the dropdown menu is a date input field with a calendar icon on the left. To its right is the label 'Date of Death'.

Figure 7: The page for migration and death in electronic registry for patients with beta thalassemia major for Mazandaran province, IR Iran 2016.

Discussion

Beta thalassemia is considered a rare disease in many developed countries. Still there are national registries for monitoring the changes in epidemiology of patients. The Thalassemia Clinical Research Network (TCRN) of the National Heart, Lung, and Blood Institute examined the demography and natural history of all patients with thalassemia who are registered in the 5 largest treatment centers in North America. The goal of the TCRN is to provide information on the changing face of this disease and the implications for diagnosis, counseling, and treatment (Conte et al., 2015). There is an Italian multicentric thalassemia registry created in 182 centers through Italy. One thousand eight hundred and seventy three patients, 259 in pediatric age have been registered (Vichinsky et al., 2005).

Whenever the disease is severe enough to bring patients to medical attention, a hospital based registry is easier to establish than a population based registry. For some conditions probably a population based registry might be appropriate (Taghavi et al., 2012).

There are still an underestimation of affected patients, as some patients are still out of radar of the health system. In spite of success in prevention of birth of new affected babies, due to better management, existing patients are growing older (Kosaryan et al., 2007). Also, because of the big number of patients and great variability of their medical conditions and needs, an electronic registry was needed to have a correct and on time picture of disease in the province.

Limitations

Our limitation at this phase is a biologic bank which is a valuable resource for future researches. Also we wish to make all forms and information in English language to be useful for all researchers and we could benefit their

comments. We also wish to have data of patients under Babol University of Medical Sciences too.

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