β thalassemia major patients profile in Ninevah governorate-Iraq

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Abstract

Background: In many developing countries the haemoglobinopathies form a major genetic problem and many thalassemia persons frequently suffer through all their life and may die unnecessarily from thalassemia, in addition to the lifelong treatment costs and the invasive interventions. The objectives of this study are to highlight the impact of thalassemia major and its management in Ninevah Governorate, to analyze the cost of caring for children with β-thalassemia major in Mosul and to dig for the socio-economic consequences of this preventable health problem.
Methods: The data were obtained partly in a straight line to thalassemia center records. A cross-sectional survey of 292 β-thalassemic patient's through a direct face-to-face interview using specially designed simple open-ended questionnaire instrument.
Results: The total number of β- thalassemia patients registered in our province is 1028 patients, 50 to 76 children born as β-thalassemia per annum and average 13 deaths /year.
About 80% of them are children <15 years, relatively low socio-economic population status. 84.9% of the surveyed families had mis-knowledge or no knowledge about the disease causation, 90.4% of them did not expect the disease occurrence, in spite of 89.4% of them have heard about this disease. Marriage among relatives was prominent.
Almost one third of studied thalassemic patient's families' fall below line of poverty that is beside the terrible sociological and emotional possessions upon family.
Each thalassemia patient cost the health institution $4320055 as a total average direct medical cost / annually.
Conclusion: The majority of thalassemic patients are relatively of low socio-economic status. Most of the surveyed families were in need for authentic information about disease causation. Almost one third of thalassemic patients families in our province fall below the line of poverty. The average cost of management bypasses the cost of prevention by 196 times. All these necessitate the setting up of a supported optimum treatment, community education and routine premarital screening for thalassemia.

Keywords: thalassemia profile, socio-economic impact, premarital screening for thalassemia

Introduction

Thalassemia is one of the most common genetic disorders. It is estimated that 60,000 to 70,000 infants are born with beta thalassemia major each year worldwide especially in the Mediterranean area, Middle East, Far East, and East Asia(1). The prevalence of β-thalassemia is 9.0 per 1000 in Saudi Arabia whereas it is 0.07 per cent in the Sultanate of Oman and the overall prevalence ranges approximately from 3 to 100 patients per 100,000 people in different Iranian provinces (1-3). In a recent study from the United Kingdom, It was found that 50% of patients with thalassemia major had died before age 35(4).

β thalassemia major is associated with life-long transfusion-dependent anemia, short stature, facial abnormalities, delayed or absent puberty, and attendant stigmas and psycho-social problems(5). Its treatment consisted of blood transfusions and chelating agents to reduce iron overload and haemochromatosis، Costs of the chelating agent، desferrioxamine, in particular and thalassaemia care, in general are expensive(6).

It was deliberated to tackle this medico-social health problem, to highlight the impact of thalassemia and its management in Ninevah Governorate, to analyze the cost of caring for children with β-thalassemia in Mosul; and to dig for the socio-economic consequences of this health problem.

Patients and methods

The study was conducted in Mosul city-Nineveh governorate /Iraq, during the period from 1st of February 2011 to 31st of June 2011. The study was conducted in thalassemia center which was established in 8/1/1997within Ibn Al-Atheer pediatric referral hospital.

To assess the completeness, the time needed and the suitable modification required for the questionnaire form, moreover to test the cooperation and the difficulties which were likely to be faced, a pilot sample consisted of 20 thalassemia patients was taken from the same center, This sample was excluded from the study target. Thalassaemic patient's profile survey was performed, following the obtaining of the official approvals from Nineveh Directorate of health.

The study population includes 292 β-thalassemia patients who attend the mentioned center during the period of study. The data were obtained partly from thalassemia center records, in addition to a direct face-to-face interview with the patients or their parents/ relatives who accompanied them. All the dialogues were completed by the same specialist. After taking a verbal consent from each respondent (or his/her relative); data collection was covered via a specially designed simple open-ended questionnaire instrument that contains (in addition to the general demographic information), social information, questions about the management, quality of life, experiences, any emotional conflicts or concerns the patients may have, direct medical and non-medical cost determination as well as indirect cost by inquiring the patients, their relatives and the health workers.

Absolute, relative and cumulative frequencies, mean and standard deviation, were computed. Data was analyzed using Statistical Package for Social Sciences (SPSS Inc, version 11Chicago, IL, 1999).

Results

The total number of β- thalassemia patients registered in Ibn Al-Atheer thalassemia center was 1028 patients with an average monthly thalassemia center visits for treatment as outpatients is 630.The point prevalence in our province is 10.3 per 10000 among less than 15

years aged child, with an absolute numbers of 50-76 β- thalassemic child born per annum, and average 13 deaths /year.

An average expenditure of 2 blood bags and 29 deferoxamine vials /patient/month was required among studied sample.

The demographic characteristic of the study sample is shown in table (1), it reveals that about four-fifth of the sample is children below 15 years. The majority of thalassemia sample (69.9%) is from urban roots, 92.8% of thalassemia patient's mothers are housewives, and 70% of the fathers are private employee. The monthly income among more than half of the sample is about $200-400, less than $200 in 31.8% and $400 and above in 16.2% . Four fifths (80.5%) of the patients' families comprise of 5 members and more, with a potentially low parents educational level.

Table (2) depicted that 89.4% of study participants have heard about thalassemia, and (90.4%) of them did not expect disease occurrence. The predominant part of the sample (60.9%) does not know the cause of thalassemia, and one-fifth of them blame the mothers as a cause of the disease according to their understanding.

Table (3) demonstrates the parent's consanguinity characteristics. The parents are relatives in 72.3% of the sample and close relative (cousins) in 65.1%. The table also shows that 39.7 of the families have one thalassemic child, 41.5% have two thalassemic patients and 18.8% have three thalassemic children and above.

The economic impact of thalassemia on the patient and Thalassemia Center within Ibn Al-Atheer Hospital is revealed in table (4). The cost were converted from Iraqi Dinars to US dollars at the rate of 1200 dinars per each US$. On the topic of patient or family expenses, this table depicts that the total average indirect management cost for each thalassemia patient per month is $67, including the monthly total average transport cost for blood transfusion visit, appliances cost needed for receiving iron-chelating drugs (scalp vein canulae ,syringes, distal water), additional monthly cost of managing other additional complications, infections and for buying gifts to release patient tension during and after medical interventions . The additional payments the family may incur on; splenectomy, treatment of its allied surgical complications, cost of probably needed cholycystectomy and expenses of bone marrow transplantation (if the patient got such a chance outside the country on the alimony of governmental or non-governmental organization) collectively for each patient is $1976.

Table (4) also explicated that each thalassemia patient cost the health institution $4320055 as a total average direct medical cost /patient annually, counting the deferoxamine vials annual cost / pateint, blood transfusion bags cost and follow-up investigations (heamoglobin level, serum iron ,serum transferrin, blood sugar ,renal and liver function tests).Diagnostic (heamoglobin vareint test), deferoxamine infusion apparatus. If the patient acquired the opportunity of bone marrow transplantation, this will incurred hospital additional $150000, rendering the total average cost for each patient to be $150885. The same table shows that the total management cost for all patients is $155109780 while the average number of couples intended to marriage yearly is 39485 rendering the total cost for prevention is $789700 taking in consideration that the cost of hemoglobin variant test for couple is $20, this illuminate that the treatment cost is 196 times than the prevention one.

Table (5) indicates that the management of thalassemia affects the pragmatic life of the family through increasing sick-leaves in 95% of those who are governmental employees. Brushing aside

the private work in 90.3% of them. In addition to repeated school absenteeism along with the poor school performance in 100% of thalassemia school students seen in the study sample, 22 of those children did not attend school from the beginning (not tabulated).

About two-thirds of thalassemia patient's mothers seen in this study experienced various sociological harassment, as table (5) showed that about one-quarter of those mothers reported to be threatened for divorce, 24.2% were threatened for husband's remarriage, and exposure to various types of violence by husband in 10.5% of the mothers.

The study also anticipated that thalassemia forms a life-size problem among the mothers and the patients simultaneously, expressed itself through emotional conflict as sorrow, and feeling of guilt which was found in more than two-thirds of the mothers, adding to the feeling of stress and frustration that experienced in more than three-quarters of the thalassemia patients who can express their feelings (Table 5).

Discussion

This study found that there were 50-76 new β- thalassemia cases born annually. In Dohuk the estimated number of affected children with a major haemoglobinopathy was 39 per year (8). This difference can be explained by variation in population density.

Point prevalence of 10.3 per 10000 < 15 yrs age child was found in this study whereas in Dohuk and Erbil with a population of around 2.2 million had more than 700 registered transfusion-dependent thalassemia patients in the two provinces(8). In general the difference in regional prevalence may be attributed to the variation in carrier rate and the frequency of consanguineous marriage. During a 5-year period in Shiraz, southern Iran,65 thalassemia patients with a mean age of 16.1 ± 4.2 years, died (9), a result which is similar to our finding of 13 thalassaemic patients' death /year.

The high rate of potentially low parental educational levels and relatively low socio-economic status seen among thalassemic families in this study was also evident in other thalassemic territories like Pakistan where majority of parents of thalassaemic children in Karachi were of low socioeconomic class and 66.7% were illiterate (10). This may explain the surprising rate of mis-knowledge or even no knowledge about the disease causation among 89.4% of the surveyed families. Despite that 89.4% of respondents have heard about thalassemia among their relatives and surroundings, 90.4% of them did not expect to face this problem among their siblings. Likewise majority of parents of Karachi thalassaemic children did not know that thalassemia is an inherited disorder (11). Subsequently, public awareness with information needed to make scope of educated decisions about thalassemia prevention and treatment should be broadened.

Marriage among relatives was a prominent consanguinity character seen among 72.3% of our thalassemia sample. Similarly high frequency of consanguineous marriages (exceeding 55%) is found in Saudi Arabia a region in which β-thalassemia carrier rate is 1.8% (2), This type of marriage ,the frequency of thalassemia trait in the population as well as the large family size, and the population density differences contribute to the our finding of frequent new β- thalassemia cases born annually in our locality considering that thalassemia is inherited as autosomal recessive disease. In a recently conducted three months lasted study in Ninevah province during 2011 revealed that 18(3.9%) out of 465 tested couples had thalassemia trait (11). The prevalence of

β-thalassemia in Saudi Arabia steadily decreased from 32.9 to 9.0 per 1000 examined persons after six years of premarital screening (2).

This study displays that, the total average management cost including the direct non-medical and indirect cost for each thalassemia patient is $67 per month. In contrast the monthly income of 83.8% of the studied families was range between <$200-$400. This means that each single patient management expense about 1/3 to 1/6 of the total family income. Taking in consideration that more than half of these families have 2-3 thalassemic patients, in this condition we can predict that one to two-third to total monthly income of more than third of these families spent on the management of these patients. That is beside other $ 1976the families may pay out for expenses of splenectomy, bone-marrow transplantation and the related complications. The last intervention may cost the institution up to $150000. A lower indirect monthly cost observed in Thailand where the average annual cost per thalassemic patient was US$950 during the year 2005; 24% of which was indirect cost comprises $ 19 for each patient per month (12), which may be ascribed to difference in standard of living, time of study conduction, quality and cost of the needed indirect services between the two regions. In addition to that, 80.5% of our study families are large families' ≥5 members. Subsequently, the treatment of β thalassemia patients in our locality could render almost one third of the families to reach the line of poverty.

The annual average direct medical institution's expenses for each thalassemia patient is $4320055 counting the yearly expenses on; investigations, deferoxamine infusion, blood transfusion, excluding other expenses like thalassemia center operating cost as salaries, equipments, and other materials. The average direct cost of each thalssemic patient in this study was $150885.This expenditure is more than that in Thailand where the average cost of thalassemia patients at the teaching hospitals was US$1,297 (13). This may attributed to the differences in the grade of patient's severity.

This study found that β thalassemia management cost incurred by health institution in Ninevah province is $4441016540 ($4320055×1028) annually. Moreover, if presumptive bone-marrow transplantation is eventuated for a patient this will exacerbate the institutional cost to be $150885 for each patient and for all thalassemic patients will be $155109780. In the same topic, this study clarify that the average cost of management bypasses the cost of prevention by 196 times. This is in assertion with the fact that the investment in prevention is worthier than treatment speculation. In Iran It is estimated that approximately 70 million dollars annually are spent for the treatment of this disease (14). According to Payne et al the yearly cost of infused iron chelation therapy in the United States is $34,460.00 for the initial treatment year and $30,004.00 for subsequent years (15).

This study illustrated that the economic effects of thalassemia on families is not through direct cost only but indirectly through its lion's share effects on fathers, mothers occupational commitment, time continuity(numerous leaves), and accuracy(work ignorance). At the same time, this disease shows its dreadful effects on patient's school performance as other chronic diseases owing to recurring hospital visits and school absenteeism turn such child to perform poorly or to dropout before graduation. Off course, this may associated with future unemployment, mal adaptive behavior, wasted opportunities and welfare costs. Moreover, β- thalassemia as a distressing disorder is found to be a feracious ground of a terrible sociological and emotional

possessions upon family milieu by mounting two-third of the patient's mothers subjected to be threatened for divorce, husband's remarrying and women disempowerment. This habitually reinforce low self-esteem that prevail among those mothers and make 68.2% of them feel sorrow and guilty. Along with feeling of frustration and stress that experienced among 76.4% of the patients, as a consequences of long course treatment, aggressive intervention, unfortunate quality of life. Comparable results found in Turkish study (16). Efforts, therefore, need to be directed to sensitize the communal consciousness regarding prevention of the disease, and towards premarital screening for detection of carrier status. Such steps are urgently needed in Iraq, in order to curtail the disease and will help to alleviate the gloomy socio-economic burden of thalassemia, and released resources for the optimum care of affected individuals.

Conclusions:

The majority of thalassemic patients are with relatively of low socio-economic status. Most of the surveyed families had mis-knowledge or no knowledge about the disease causation. In 90.4% of those families they did not expect the disease occurrence. Marriage among relatives was prominent. Almost one third of studied thalassemic families fall below the line of poverty in our province. This study recommends the routine thalassemia premarital screening as worthy preventive approach in Ninevah governorate and the need of governmental commitment to support the affected families.

References

1-Abolghasemi H, Amid A, Zeinali S et al. Thalassemia in Iran: epidemiology, prevention, and management. J Pediatr Hematol Oncol. 2007 Apr; 29(4):233-238.

2-Memish ZA, Saeedi MY. Six-year outcome of the national premarital screening and genetic counseling program for sickle cell disease and β-thalassemia in Saudi Arabia. Ann Saudi Med. 2011 May-Jun; 31(3):229-235.

3-Al-Riyami A, Ebrahim GJ. Genetic blood disorders survey in the Sultanate of Oman. J Trop Pediatr. 2003 Jul; 49:1-20.

4-Borgna-Pignatti C, Cappellini MD, De Stefano P et al. Survival and complications in thalassemia. Ann N Y Acad Sci. 2005;1054:40-47.

5-Politis C. The psychosocial impact of chronic illness. Ann NY Acad Sci 1998; 850:349.

6-Ginsberg G, Tulchinsky T, Filon D et al. Cost-benefit analysis of a national thalassaemia prevention programme in Israel. J Med Screen. 1998; 5(3): 120-126.

7-Al-Allawi NA, Al-Dousky AA. Frequency of haemoglobinopathies at premarital health screening in Dohuk, Iraq: implications for a regional prevention programme. East Mediterr Health J. 2010 Apr;16(4):381-385.

8-Al-Allawi NA, Hassan KM, Sheikha AK et al. β-Thalassemia Mutations among Transfusion-Dependent Thalassemia Major Patients in Northern Iraq. Mol Biol Int. 2010:4.

9-Bazrgar M, Peiravian F, Abedpour F, Karimi M. Causes for hospitalization and death in Iranian patients with β-thalassemia major. Pediatr Hematol Oncol. 2011 Mar; 28(2):134-139.

10-Arif F, Fayyaz J, Hamid A. Awareness among parents of children with thalassemia major. J Pak Med Assoc. 2008 Nov; 58(11):621-4.

11-Mohammed F.F,Fadhil N.A,Ballo A. Premarital diagnosis of hemoglobinopathies in Mosul. Program and abstract of 9th Mosul medical conference organized by ministry of health, directorate of health in Ninawa 2011:44.

12-Riewpaiboon A, Nuchprayoon I, Torcharus K et al. Economic burden of beta-thalassemia/Hb E and beta-thalassemia major in Thai children. BMC Res Notes. 2010 Jan 30; 3:29.
13-Goljan. E. Pathology. 2nd ed. Mosby Elsevier Rapid Review Series 2006.
14-Iran T.A. A report of Iranian children and young adults afflicted with β-thalassemia major. Iranian society of thalassemia 2006; 11: 24-26.
15-Payne KA, Desrosiers MP, Caro JJ et al. Clinical and economic burden of infused iron chelation therapy in the United States. Transfusion 2007; 47(10):1820-1829.
16-Canatan D, Ratip S, Kaptan S, Cosan R. Psychosocial burden of beta-thalassaemia major in Antalya, south Turkey. Soc Sci Med. 2003 Feb; 56(4):815-819.

Table (1): Demographic characteristics of the sample

	N= 292	
Age in years	**No.**	**%**
1-5	89	30.5
6-10	71	24.3
10-15	68	23.3
16-20	39	13.3
> 20	25	8.6
Residence		
Rural	88	30.1
Urban	204	69.9
Mother's occupation		
Housewife	271	92.8
Governmental employee	18	6.2
Private works	3	1.0
Father's occupation	**No.**	**%**
Governmental employee	48	16.2
Private employee	206	70.6
Military	5	1.8
Un-employed	21	7.2
Retired	6	2.1
Others*	6	2.1
Monthly income:	**No.**	**%**
<$200	93	31.8
$200-400	152	52.0
$400-600	36	12.4
>$600	11	3.8
Family no.	**No.**	%
<5	57	19.5
5-7	114	39.1
>7	121	41.4
Education	**Mean + SD**	
Mother's years of education	5.3± 3.9 years	
Father's years of education	8.0 ± 4.6 years	

*Others include: the disabled, the dead & the students

Table 2: Parent and patient knowledge about thalassemia

	N=292	
Previous disease knowledge	**No.**	**%**
Yes	261	89.4
No	31	10.6
Did they expect disease occurrence	**No.**	**%**
Yes	28	9.6
No	264	90.4
Knowledge of disease causation	**No.**	**%**
Father is the cause	11	3.8
Mother is the cause	59	20.2
Both are the cause	44	15.1
Don't know	178	60.9

Table 3: Parent Consanguinity of the sample

	N=292	
parent are relatives	**No.**	**%**
Yes	220	75.3
No	72	24.7
Degree of parent's relationship	**No.**	**%**
Close relative	190	65.1
Far relative*	30	10.2
Not relative	72	24.7
Number of patients / family	**No.**	**%**
1	116	39.7
2	121	41.5
3	34	11.6
>4	21	7.2

Table 4: Average cost of disease related expenses

Patient's responsibilities	Average no./patient/ month	Average cost/event/patient ($)	Total average Cost/patient ($)
Cost of blood transfusion visit/month	2	10	20
Cost of appliances needed for receiving deferoxamine/month	29	1	29
Additional monthly cost	NA*	18	18

			67
Average cost of Possibly needed Interventions**			
Splenectomy	NA	416	416
Bone marrow transplantation	NA	1500	1500
Cost of surgical complications	NA	60	60

			1976
Average Institution's medical responsibilities	Average no./family	Cost/ each event ($)	Total average cost/patient ($)§
Average deferoxamine vial expense/patient/year	NA	10	3480000
Blood bag expense/patient /year	NA	35	840000
Various blood tests /patient/year	NA	NA	55
Total average direct cost/patient/year			4320055
Hemoglobin Variant test/pateint	4	10	40
Average deferoxamine vial expense/patient	NA	10	10
Blood bag expense/patient	NA	35	35
Deferoxamine infusion apparatus	NA	800	٨٠٠
Bone marrow transplantation cost	NA	150000	150000
Total average cost /each patient	NA	NA	150885
Total cost of treatment for all patients	NA	**NA**	155109780
Total cost for prevention	NA	**NA**	789700

*NA: Not applicable

** : From personal interview with companion of patients subjected to such events.

§: Data obtained from thalassemia center record.

Table 5: Effects of thalassemia on socio-economic status of family

Effect on father's and patient's work	Yes	Total	%
Governmental employee	46	48	95.8
Private employee	186	206	90.3
School performance	164	164	100.0
Effect on family life			
Threat by divorce	75	292	24.9
Threat by abandon	8	292	2.9
Threat of husband's remarriage	70	292	24.2
Exposure to violence	30	292	10.5
No threats	109	292	37.5
Effect on emotional aspect			
Sorrow, guilty (mothers)	199	292	68.2
Frustration, stress (patients)	223	292	76.4

لمحة عن حياة مرضى الثلاسيميا في محافظة نينوى

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خلفية البحث: تشكل اعتلالات الهيموكلوبين مشكلة وراثية رئيسية في العالم و تتسبب هذه الأعتلالات في الكثيرمن المعانات بالنسبة لمرضى الثلاسيميا خلال حياتهم والتي قَدْ تنتهي بالموت هذا بالأضافة إلى كلفَ المعالجةِ الدائمةِ والتداخّلاتِ المؤلمة.اهداف البحث هي لإبْراز الأعباء المترتبة على علاج مرض الثلاسيميا في محافظةِ نينوى، ولتَحليل كلفةِ العناية بأطفالِ الثلاسيميا، ولأيضاح النتائج الإقتصاديةِ والإجتماعيةِ لهذه المشكلةِ الصحيةِ التي بالأمكان منعها.
طريقة البحث:استندت الدراسة جزئيا على سجلات مركز الثلاسيميا في المحافظة، هذا بجانب دراسة المقطع العرضي لعينة متكونة من ٢٩٢ مريض مصاب بالبيتا ثلاسيميا من خلال نموذج استبياني خاص، بسيط ومتكامل.
النتائج: أوضحت الدراسة أن هناك ١٠٢٨ مريض بالبيتا ثلاسيميا في محافظة نينوى. وأن ٥٠-٧٦ طفل يولدون سنويا بهذا المرض وبمعدل ١٣ وفاة في المحافظة سنويا. ٨٠% منهم أطفال دون الخامسة عشر من آسر ذات مستويات اقتصادية وثقافية متدنية نسبيا، ثلثهم يقع تحت خط الفقر، ٨٤.٩% منهم ليس لديهم فكرة عن سبب المرض، ٩٠.٤% لم يتوقعوا حدوثه لدى أطفالهم رغم سماعهم به من أقاربهم ، زواج الأقارب كان سمة غالبة لديهم. كما أوضحت الدراسة الأبعاد الأجتماعية والعاطفية للمرض على المريض والعائلة. وأن مريض الثلاسيميا الواحد يكلف المؤسسة الصحية 4320055$ كمعدل عام للمصروفات العلاجية المباشرة سنويا.
الأستنتاجات: غالبية مرضى البيتا ثلاسيميا في محافظة نينوى هم من أسر ذات مستويات أجتماعية وأقتصادية متدنية، ثلثهم يقع تحت خط الفقر، تنقصهم المعرفة بأسباب المرض، وزواج الأقارب سمة واضحة لديهم. ان متوسط كلفة العلاج تتجاوز كلفة الوقاية ب 196 مرة. كل هذا يستلزم تبني تدابير علاجية ناجعة وحديثة،هذا بجانب التثقيف المجتمعي والفحص الروتيني قبل الزواج عن مرض الثلاسيميا لمنع حدوث المرض.