Thalassemia congress

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Original Article

Treatment Status of Patients with B-Thalassemia Major in Northern Iran: Thalassemia Registry System

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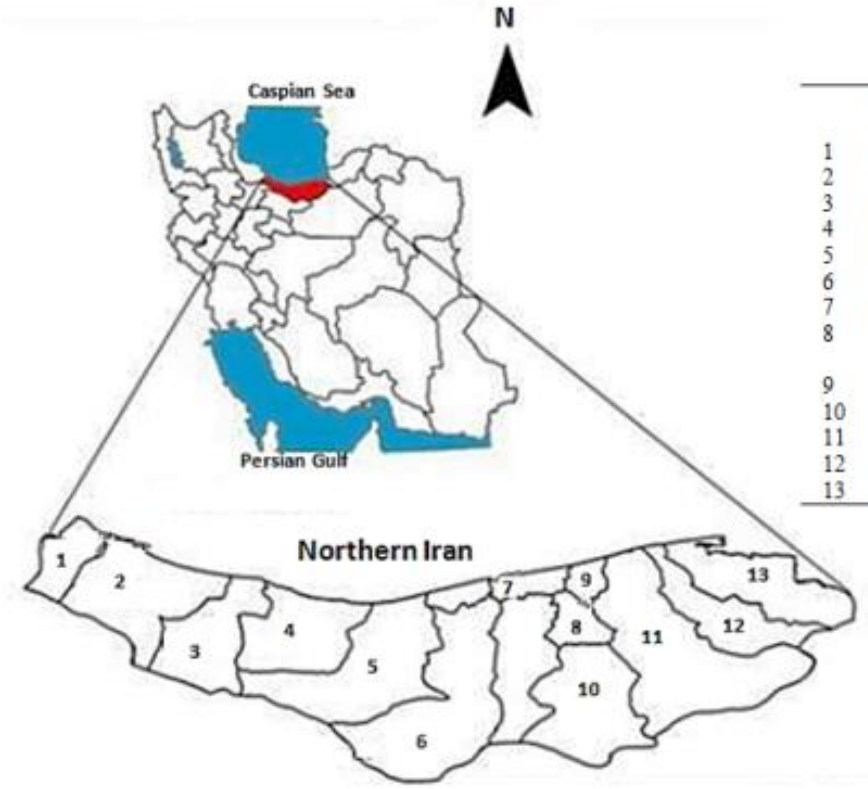
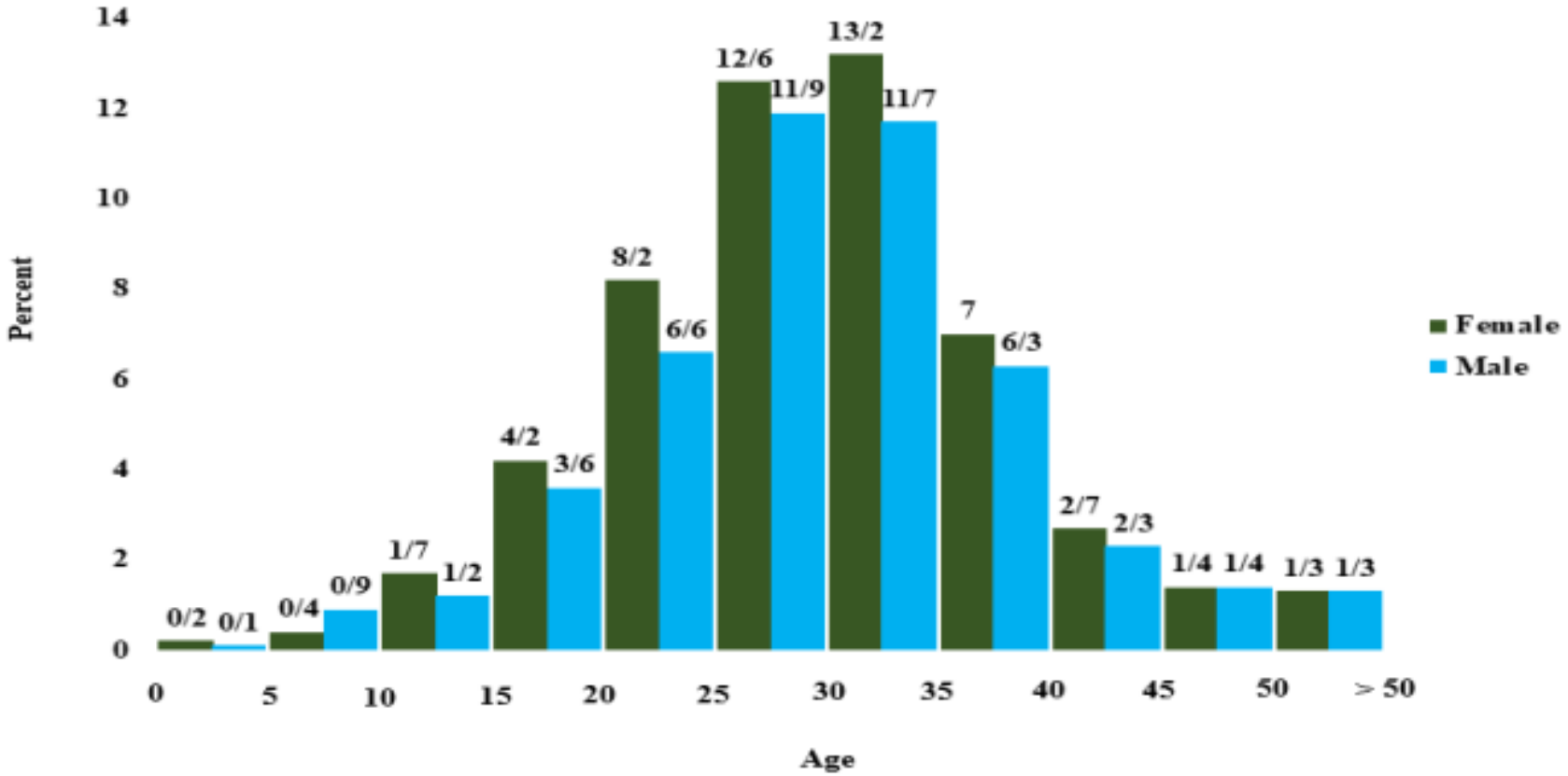
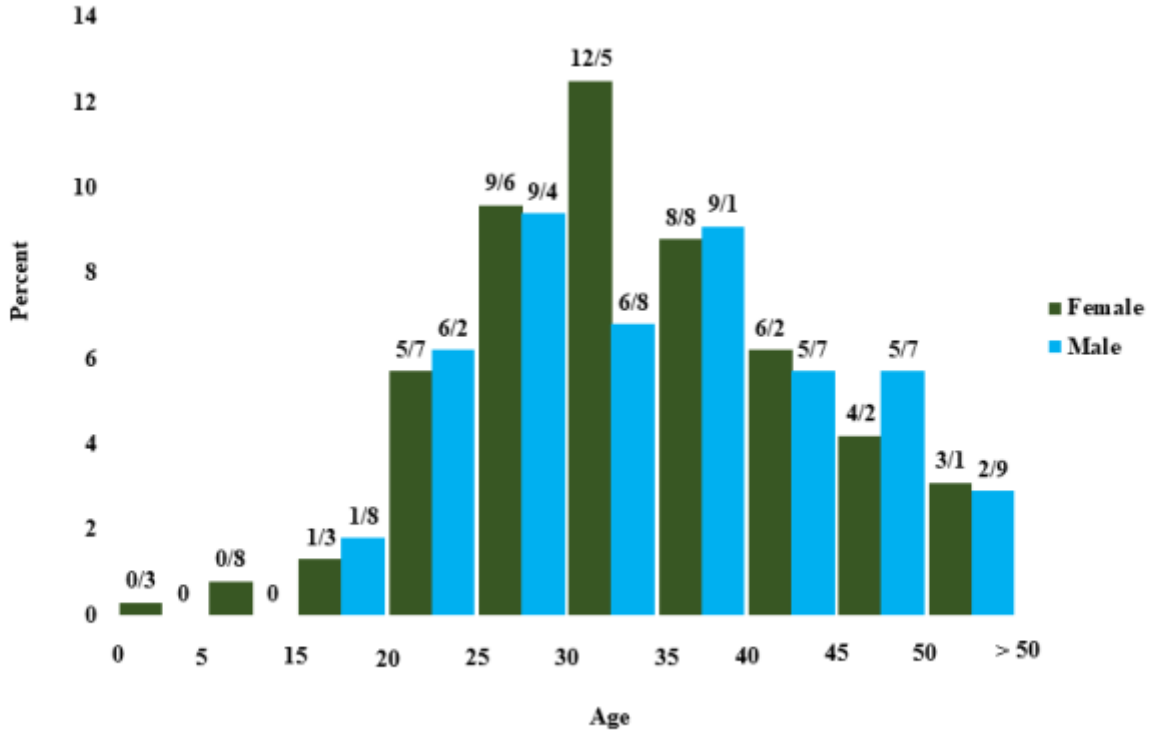
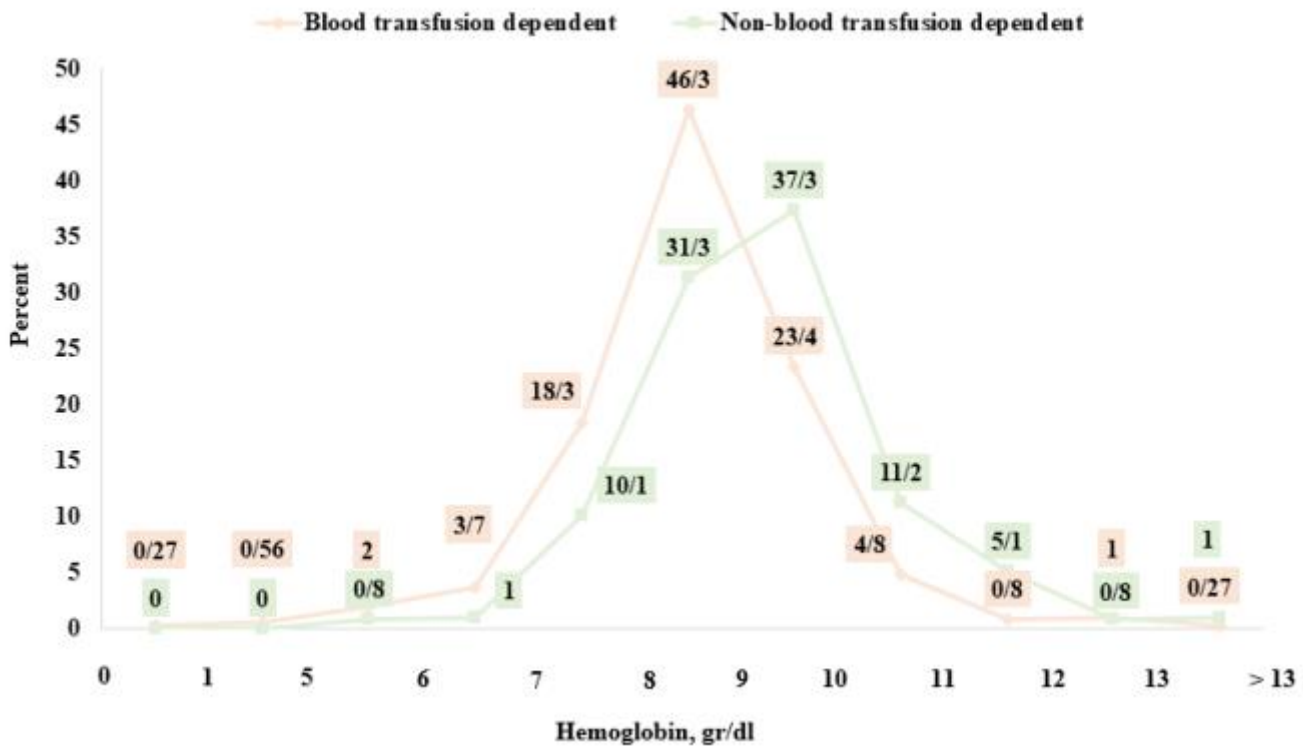


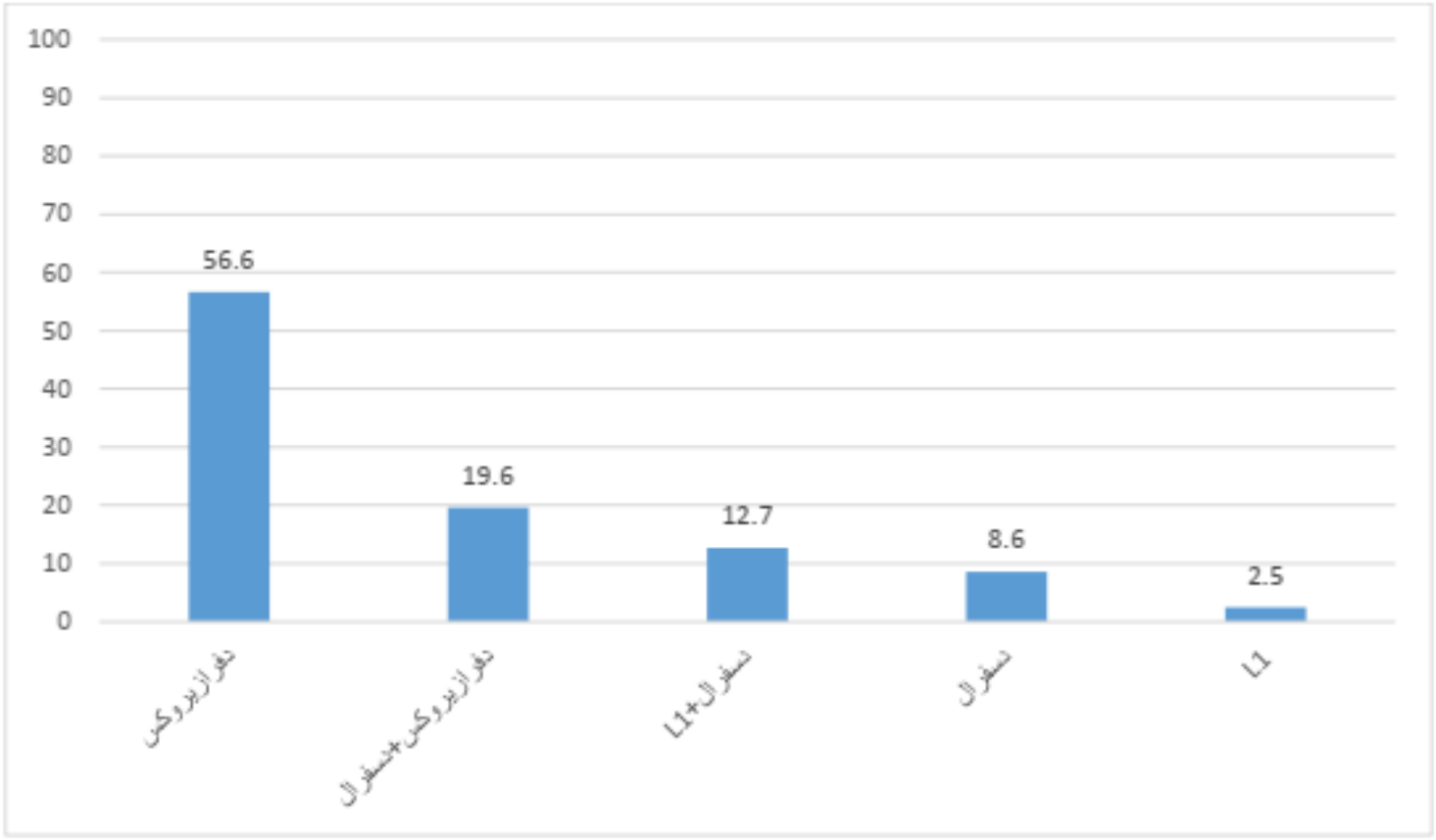
	City	Hospital name	Number of patients
1	Ramsar	Imam Sajjad Hospital	37
2	Tonekabon	Shahid Rajaee Hospital	98
3	Chalus	Ayatollah Taleghani Hospital	49
4	Noshahr	Shahid Beheshti Hospital	21
5	Noor	Imam Khomeini Hospital	66
6	Amol	Imam Reza Hospital	408
7	Fereydunkenar	Imam Khomeini Hospital	83
8	Ghaemshahr	Razi Hospital	171
		Valiasr Hospital	61
9	Joybar	Azizi Hospital	31
10	Zirab	Shohada Hospital	11
11	Sari	Bu-Ali Sina Hospital	615
12	Neka	Thalassemia Clinic	63
13	Behshahr	Imam Khomeini Hospital	91


14
12
10
8
6
4
2
0
Percent
0/2
0/1
0/4
0/9
1/7
1/2
4/2
3/6
8/2
6/6
12/6
11/9
13/2
11/7
7
6/3
2/7
2/3
1/4
1/4
1/3
1/3
0
5
10
15
20
25
30
35
40
45
50
> 50
Age
Female
Male

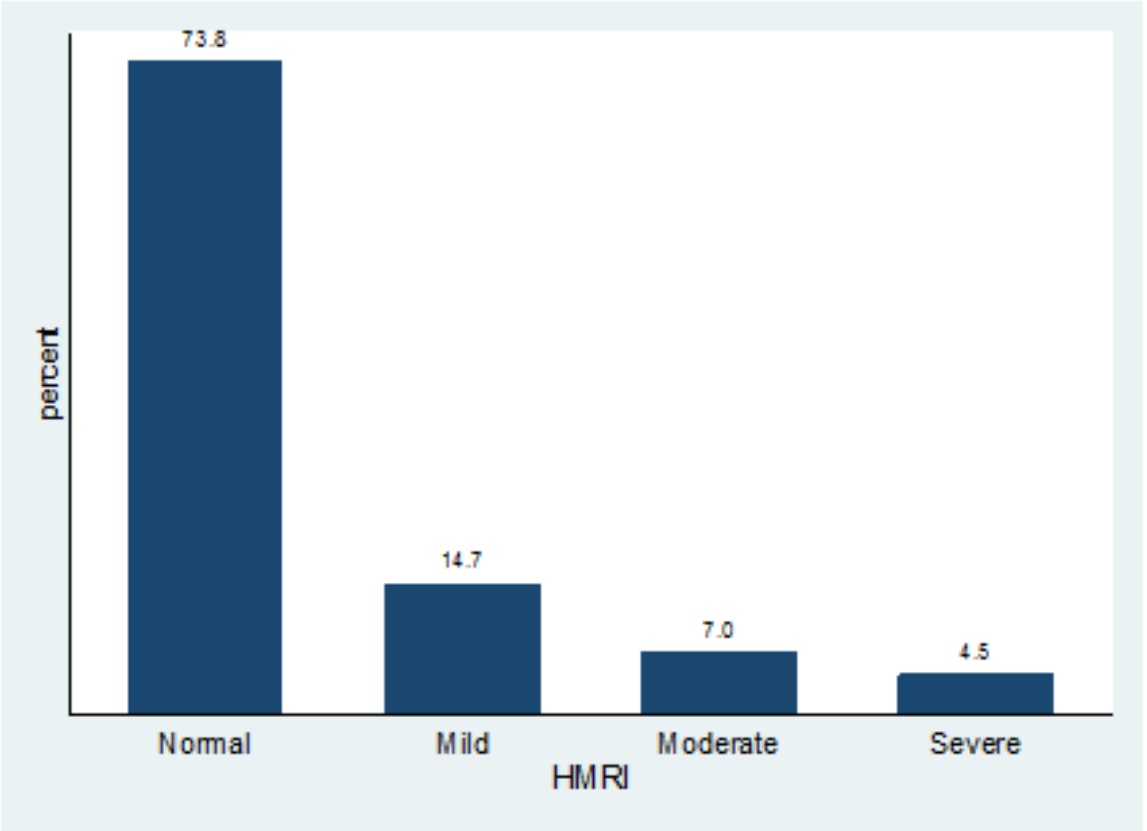


: Distribution of 446 transfusion-dependent patients according to age and gender, Mazandaran Province,

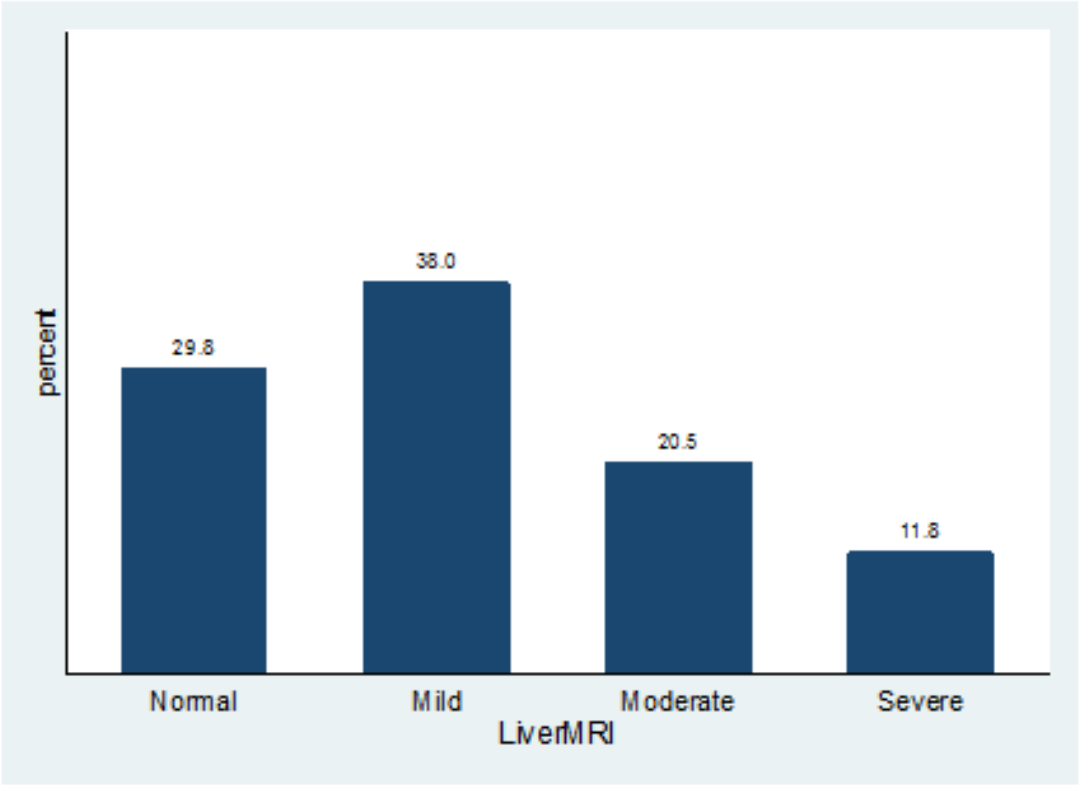

Blood transfusion dependent
Non-blood transfusion dependent
Percent
50
45
40
35
30
25
20
15
10
5
0
0
1
5
6
7
8
9
10
11
12
13
> 13
Hemoglobin, gr/dl
0/27
0/56
2
3/7
18/3
46/3
23/4
4/8
5/1
1
1
0
0
0/8
1
10/1
31/3
37/3
11/2
0/8
0/8
0/27



درصد بیماران وابسته به تزریق خون تحت منوتراپی و درمان ترکیبی با داروهای آهن زدا– بیمارستان بوعلی ساری - 1400 (n= 408)



HMRI	Freq.	Percent	Cum.
Normal	296	73.82	73.82
Mild	59	14.71	88.53
Moderate	28	6.98	95.51
Severe	18	4.49	100.00
Total	401	100.00	



LiverMRI	Freq.	Percent	Cum.
Normal	119	29.75	29.75
Mild	152	38.00	67.75
Moderate	82	20.50	88.25
Severe	47	11.75	100.00
Total	400	100.00	

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ORIGINAL PAPER

Therapy area: Other

A double-blind, controlled, crossover trial of amlodipine on iron overload status in transfusion dependent β-thalassemia patients

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Abstract

Background and aim: This study examined whether administration of amlodipine could improve myocardial iron loading status in patients with transfusion dependent β-thalassemia (TDT), through a placebo-controlled, crossover study.

Methods: Amlodipine (5 mg, daily) or placebo were prescribed to all patients (n = 19) for 6 months, and after a 2-week washout period, patients were crossed over to the other group. The efficacy of amlodipine on iron loading was assessed by measuring myocardial T2*-weighted magnetic resonance imaging (MRI T2*, millisecond [ms]) and serum ferritin (ng/mL).

Results: Seventeen patients completed the study. The mean ± standard deviation [SD] of myocardial MRI T2* at baseline was 9.83 ± 2.67 ms Myocardial MRI T2* value rose to 11.44 ± 4.14 ms post amlodipine treatment in all patients. After placebo, myocardial MRI T2* value reached 10.29 ± 4.01 ms After controlling the baseline measures, Hedges's g for ferritin and myocardial MRI T2* outcomes were estimated 3.84 (95% confidence interval [CI] 2.68 to 4.97) and −1.80 (95% CI −2.58 to −0.10), respectively.

Conclusion: Amlodipine might improve myocardial MRI T2* and serum ferritin level compared to placebo. However, larger clinical studies are needed to confirm the results.

Short Communications

Calcium Channel Blockers in Conjunction with Standard Iron-Chelating Agents for β-Thalassemia Major: Systematic Literature Search

Mohammed A. Alali, Khalid M.A. Alanazi, Sarah N. Alsayil, Zakaria Omari & Ali Shaaban

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Abstract

Thalassemia is a genetic mutation of the α- or β-globin chains that lead to defective erythropoiesis. This study aimed to collect evidences from all published studies that investigated the clinical effectiveness of calcium channel blockers (CCBs) in conjunction with chelation therapy for reducing iron overload in patients with thalassemia. A systematic search was conducted in PubMed, Institute for Scientific Information (ISI) Web of Science, Scopus, Cochrane Central Register of Controlled Trials, and Virtual Health Library. Original studies reporting the use of CCBs in patients with thalassemia were included for meta-analysis. A total of five randomized studies including 210 patients were included with a follow-up period of 3–12 months. There was no significant difference between amlodipine and control groups in increasing the heart T2* magnetic resonance imaging (MRI) [mean difference (MD) 95% confidence interval (95% CI) = −1.9 (−4.4 to 0.5), $p = 0.119$] or reducing the liver iron concentration [MD 95% CI = −0.046 (−0.325 to 0.2), $p = 0.746$]. Although there were no serious adverse events reported in the included trials, further studies are recommended to strengthen our findings.

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SYSTEMATIC REVIEW & META-ANALYSIS

Coadministration of silymarin with iron chelators in transfusion-dependent β-thalassemia patients: a systematic review and meta-analysis for effect on iron overload

Hadi Darvishi-Khezri[a], Mohammad Naderisorki[b], Mohammad Zahedi[c], Parham Mortazavi[d], Fatemeh Tajik[e], Amirreza Nasirzadeh[f] and Hossein Karami[b]

ABSTRACT

Background and aim: We conducted a systematic review to apprise the efficacy of silymarin in conjunction with standard iron chelators on iron overload for transfusion-dependent β-thalassemia (TDT) patients.

Methods: We searched PubMed, Web of Science, Scopus, Sciencedirect, the Cochrane Library (the Cochrane Database of Systematic Reviews, and the Cochrane Central Register of Controlled Trials (CENTRAL) to 1 May 2020. All randomized controlled trials (RCTs) studies comparing the effect of iron chelators alone versus silymarin plus standard routine treatment on iron burden amid TDT were included in this review. Primary outcomes comprised serum ferritin level (ng/mL), liver iron concentration (LIC Fe/kg dry weight), and total iron binding capacity (TIBC mcg/dL)

Results: Combination therapy of silymarin and iron chelators showed a significant improvement in serum ferritin level in TDT patients, compared to nonsilymarin users [eight studies, n = 477]; weighted mean difference (WMD) −1.79, 95% confidence interval [CI] −2.86 to −0.72, I^2 96.1%; P = 0.001. Concurrent treatment with silymarin failed to significantly decrease LIC in TDT patients [two studies, n = 106]; WMD 0.74, 95% CI −1.62 to 3.10, I^2 96.6%; P = 0.54.

Conclusion: There is no evidence of the effectiveness of adding silymarin to standard iron chelators to reduce iron load in TDT.

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journal homepage: www.elsevier.com/locate/ctim

The impact of silymarin on antioxidant and oxidative status in patients with β-thalassemia major: A crossover, randomized controlled trial

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ABSTRACT

Background & aims: Blood transfusion therapy is lifesaving for individuals with β-thalassemia major (β-TM). Iron burden following blood transfusion is the main cause of oxidative stress (OS) and organ dysfunction in these patients. The aim of this study was to evaluate the effects of silymarin on serum antioxidant and oxidative status in patients with β-TM.

Methods: A crossover, randomized controlled trial was performed on 82 thalassemia patients. In two periods of 12 weeks, patients received 420 mg silymarin (divided into three equal 140-mg daily doses) and placebo. The washout period between the two phases was two weeks. Serum malondialdehyde (MDA), protein carbonyl (CO), total antioxidant capacity (TAC), and reduced glutathione (GSH) were measured before and after both periods.

Results: Sixty-nine patients completed the study. Mean serum MDA and protein CO significantly decreased in all patients with β-TM after three months of treatment with silymarin. At the end of the study, serum MDA decreased from 20.36 ± 20.11 to 4.79 ± 4.71 μmol/l (compared to 17.81 ± 16.05 μmol/l after administration of placebo), and protein CO dropped from 0.31 ± 0.28 to 0.11 ± 0.09 mM/l (compared to 0.24 ± 0.17 mM/l with placebo). Additional laboratory parameters (such as serum TAC and plasma GSH) were also significantly elevated after therapy with silymarin. At the end of the study, serum TAC increased in all patients from 620.7 ± 202.64 to 971.83 ± 328.16 μmol $FeSO_4$/l (compared to 672.22 ± 206.88 μmol $FeSO_4$/l with placebo), and GSH increased from 46.16 ± 41.68 to 195.35 ± 210.98 nM/l (compared to 58.52 ± 48.95 nM/l with placebo). The treatment effect of silymarin was measured using a mixed-effects model of variance analysis for changes in MDA, protein CO, TAC, and GSH, with significant effects being demonstrated for each laboratory parameter ($P < 0.001$, $P = 0.002$, $P < 0.001$, and $P < 0.001$, respectively).

Conclusions: Silymarin was effective in decreasing serum OS and enhancing serum antioxidant capability in patients with β-thalassemia major. Silymarin given as an adjuvant therapy to standard iron chelators may provide an improvement in the OS measurements obtained in these patients, with accompanying benefit.


Thank
you