



La terapia trasfusionale nella talassemia



Gaspare Adorno

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Policlinico Tor Vergata

La terapia trasfusionale nella talassemia

Obiettivo

Livello di emoglobina necessario per garantire
l'accrescimento e la vita di relazione

Complicanze

Sovraccarico di ferro
Immunizzazione eritrocitaria
Infezioni da HBV, HCV, HIV



La terapia trasfusionale nella talassemia



Immunizzazione eritrocitaria

alloimmunizzazione
autoimmunizzazione

La terapia trasfusionale nella talassemia

Pazienti numero	Terapia trasfusionale con fenotipi compatibili	Isoimmunizzazione %
162	ABO/Rh/K	3.7
83	ABO/D	15.7
973	ABO/D	22.6

Spanos T, Vox Sang, 1990

Table 1. Alloantibodies and RBC autoantibodies detected in thalassemia patients according to the extent of RBC phenotype of the transfused blood

	Group I (n = 29)	Group II (n = 26)	Group III (n = 35; 9 + 26)	Total no. in all patients	Total no. in Asian patients
Alloantibodies					
Anti-K*	2	4	—	6	4
Anti-c*	1	1	—	2	1
Anti-E*	1	3	—	4	1
Anti-JKb*	—	—	1	1	1
Anti-Lea	—	1	—	1	1
Anti-Kp-a*	—	1	—	1	1
Anti-M	—	1	—	1	1
Anti-i	2	—	—	2	2
HTLA	—	1	—	1	1
Total	6	12	1	19	13
Total clinically significant					
	4	9	1	14	8
Total autoantibodies					
	5	9	2	16	14

HTLA indicates high titer, low avidity.

*Indicates clinically significant.

Alloimmunization and erythrocyte autoimmunization in transfusion dependent thalassemia patients of predominantly Asian descent
ST Singer, Blood, 2000

La terapia trasfusionale nella talassemia

TABLE 1. Number of RBC alloantibodies detected in transfusion-dependent thalassemia patients according to their ethnic background

System	Alloantibodies	Kuwaiti Arab	Non-Kuwaiti Arab	Total RBC alloantibodies identified in all groups	
				Number	%
Rh	Anti-D	5	7	12	21.1
	Anti-C	4	5	9	15.8
	Anti-E	15	11	26	45.6
	Anti-c	3	2	5	8.8
	Anti-C ^w	3	2	5	8.8
	Anti-v	7	5	12	21.1
KELL	Anti-K	18	23	41	72
	Anti-Kp ^a	1	2	3	5.3
	Anti-Js ^a	1	1	2	3.5
DUFFY	Anti-Fy ^a	2	NA	2	3.5
	Anti-Fy ^b	1	1	2	3.5
KIDD	Anti-JK ^a	3	3	6	10.5
	Anti-JK ^b	1	NA	1	1.8
MNSs	Anti-S	NA	3	3	5.3
Lewis	Anti-Le ^a	6	1	7	12.3
	Anti-Le ^b	2	NA	2	3.5
	Anti-Lu ^a	1	NA	1	1.8
Lan	Anti-Lan	1	NA	1	1.8
Colton	Anti-Co ^p	1	NA	1	1.8
Diago	Anti-Dia ^b	1	NA	1	1.8
Total		76	66	142	
Total clinically significant		68	65	133	

RBC alloimmunization and autoimmunization
among transfusion – dependent Arab thalassemia patients
R.Ameen et al, Transfusion, 2003

La terapia trasfusionale nella talassemia

TABLE 2. Age range (years) at which the RBC alloantibodies were identified

Alloantibody	2-5	6-10	11-15	16-20	>20
Anti-D	5	NA	2	1	1
Anti-C	3	NA	2	NA	2
Anti-E	6	9	5	3	3
Anti-c	1	2	1	1	NA
Anti-C ^w	2	1	1	NA	NA
Anti-v	2	6	1	1	NA
Anti-K	9	11	6	5	7
Anti-Kp ^a	1	1	NA	NA	1
Anti-Js ^a	1	2	NA	NA	NA
Anti-Fy ^a	NA	1	NA	NA	1
Anti-Fy ^b	NA	NA	1	1	NA
Anti-Jk ^a	NA	2	2	1	NA
Anti-Jk ^b	NA	NA	NA	NA	1
Anti-S	NA	1	1	1	NA
Anti Lu ^a	NA	NA	1	NA	NA
Total	30	36	23	14	16

Numbers may not add up to the total due to missing value of age.

RBC alloimmunization and autoimmunization
among transfusion – dependent Arab thalassemia patients
R.Ameen et al, Transfusion, 2003

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**TRANSFUSION
MEDICINE**

Official Journal of
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British Blood
Transfusion Society

Transfusion Medicine | SHORT COMMUNICATION

Red cell alloimmunisation in regularly transfused beta thalassemia patients in Pakistan

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La terapia trasfusionale nella talassemia

2 U. Zaidi et al.

Table 1. Characteristics of thalassemia patients with alloantibodies

S. no.	Age (year)	Ethnicity	Age of first Tx (years)	Hb	Tx frequency (days)	DNA mutations	Blood group	DCT	Antibody screening	Spleen size
1	21	Sindhi	1	5.8	15	IVSI-5	A+	Neg.	Anti-E	Normal
2	6.7	Mohajir	2.5	4.8	30	IVSI-5	O+	Neg.	Anti-E	Normal
3	7.2	Pashto	2	6.6	30	IVSI-5	O+	Neg.	Anti-E	Normal
4	2.5	Mohajir	2	5.4	7	Fr 8-9	B+	Pos.	Anti-E	Enlarged
5	18	Sindhi	0.3	8.8	15	IVSI-5	O+	Neg.	Anti-K	Normal
6	13.5	Sindhi	0.7	8.8	30	IVSI-5	O+	Neg.	Anti-K	Enlarged
7	11	Sindhi	1	9.4	15	Del 619	O+	Pos.	Anti-K	Enlarged
8	4	Mohajir	1.6	6.4	15	Fr 8-9	O+	Pos.	Anti-e	Enlarged
9	5.8	Pashto	0.75	6.8	15	Fr 8-9	B+	Pos.	Anti-e	Enlarged
10	18	Sindhi	1	9	30	Del 619	O-	Neg.	Anti-D	Splenectomised
11	10.3	Sindhi	0.2	5.1	30	Cd 30	AB-	Neg.	Anti-D, -C	Enlarged
12	10	Sindhi	0.8	9	30	Del 619	A+	Neg.	Anti-E, Kell	Normal
13	5	Pashto	3	10.3	30	Cd 5	B-	Pos.	Anti-D, -E and -C	Enlarged
14	1.2	Sindhi	0.8	7.4	30	Fr 8-9	O+	Pos.	Anti-E, -K, -C, -M and Kpa	Normal

Tx, transfusion; Hb, haemoglobin.

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Frequency Of Antibodies in Alloimmunized Patients

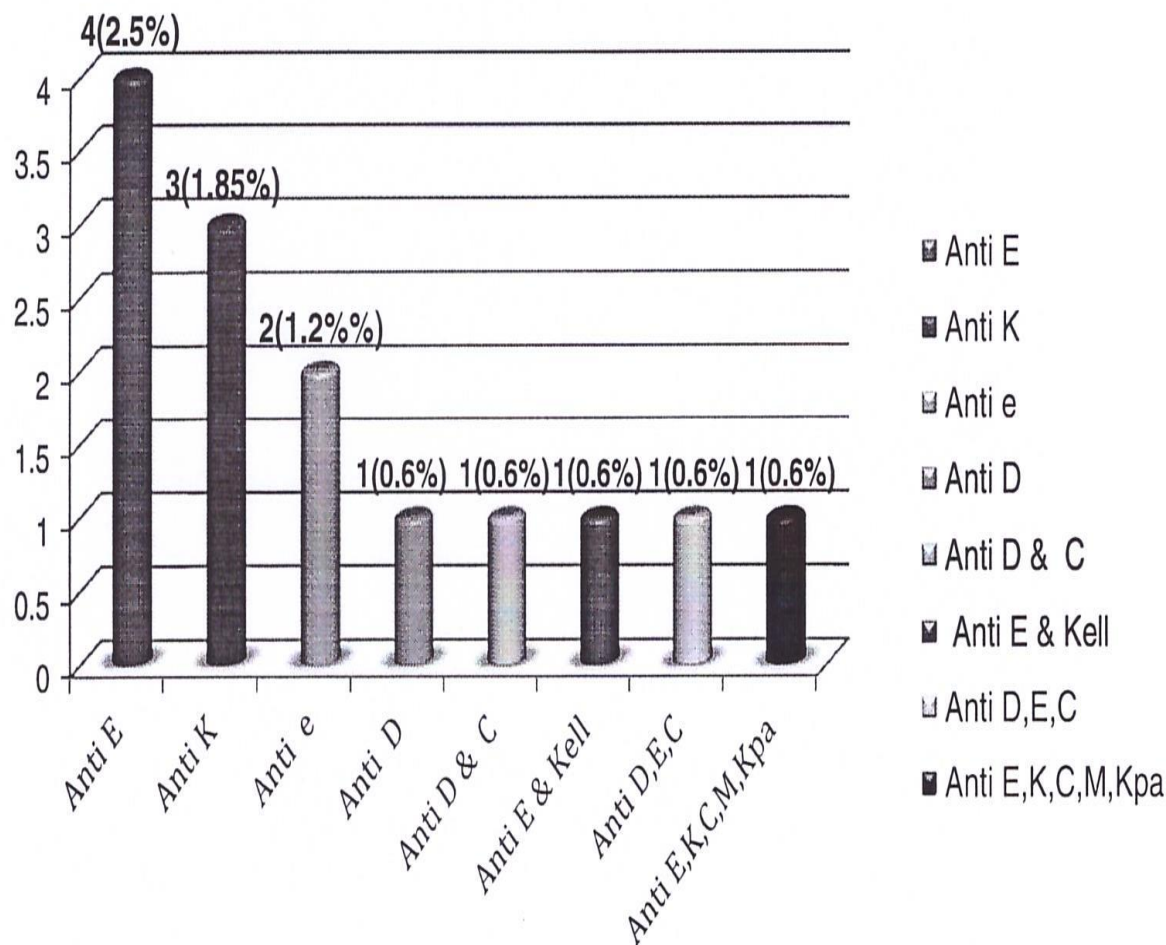


Fig. 1. Frequency of antibodies in alloimmunised patients.

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Red cell alloimmunisation 3

Table 2. Risk factors between two groups

	Without alloimmunisation 148/162 (91.4%)	With alloimmunisation 14/162 (8.6%)
Age in years (mean $\pm$ SD)	7.8 $\pm$ 5.1	9.5 $\pm$ 6.1
Age at first transfusion in year (mean $\pm$ SD)	1.2 $\pm$ 0.8	1.2 $\pm$ 0.7
Frequency of transfusion, days (mean $\pm$ SD)	31.8 $\pm$ 1	23 $\pm$ 8.8
Splenomegaly (%)	51 (34%)	7 (50%)
Post splenectomy (%)	8 (5%)	1 (7%)
Haemoglobin (mean $\pm$ SD)	7.2 $\pm$ 1.8	7.14 $\pm$ 1.8
Total leukocyte count (mean $\pm$ SD)	12.2 $\pm$ 19.3	19.5 $\pm$ 29.3
Platelets (mean $\pm$ SD)	279.5 $\pm$ 168.6	202.2 $\pm$ 94.5
DCT positive	18 (12%)	6 (43%)

La terapia trasfusionale nella talassemia



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TRANSFUSION
CLINIQUE ET BIOLOGIQUE

Transfusion Clinique et Biologique 19 (2012) 345–352

Article original

Immunisation anti-érythrocytaire dans les hémoglobinopathies : à propos de 84 cas

Red blood cell immunization in haemoglobinopathie: About 84 cases

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Disponible sur Internet le 25 octobre 2012

La terapia trasfusionale nella talassemia

I. Ben Amor et al. / Transfusion Clinique et Biologique 19 (2012) 345–352

347

Tableau 1

Caractéristiques des patients et concentrés de globules rouges transfusés.

	Patients (n = 84)	Syndromes drépanocytaires majeurs (n = 54)	Thalassémies (n = 30)
Âge moyen (<i>extrêmes</i>)	10,13 (1–45)	13,56 (1–45)	6,7 (1–28)
Sex-ratio (H/F)	1,54 (51/33)	1,57 (33/21)	1,5 (18/12)
<i>Concentrés de globules rouges totaux</i>			
Total	3545	1019	2526
Moyenne	42,2	18,87	82,4
Extrêmes	1–291	1–211	1–291
<i>Concentrés de globules rouges standard</i>			
Total	1257 (35,46 %)	335 (32,87 %)	922 (36,5 %)
Moyenne	14,96	6,20	30,73
Extrêmes	1–271	1–45	1–271
<i>Concentrés de globules rouges phénotypés (RH, KEL)</i>			
Total	2288 (64,54 %)	684 (67,12 %)	1604 (63,5 %)
Moyenne	27,23	12,67	53,47
Extrêmes	1–209	1–199	1–209

La terapia trasfusionale nella talassemia

Tableau 2

Suivi immunohématologique des patients.

	Patients (n = 84)	Syndromes drépanocytaires majeurs (n = 54)	Thalassémies (n = 30)
<i>Recherches d'agglutinines irrégulières</i>			
Total	1474	320	1154
Moyenne par patient (extrêmes)	17,5 (1-129)	5,92 (1-115)	44,38 (1-129)
<i>Tests de Coombs directs</i>			
Total	272	60	212
Moyenne par patient (extrêmes)	3,2 (1-29)	1,11 (1-24)	13,25 (1-29)

La terapia trasfusionale nella talassemia

Tableau 3
Immunisation anti-érythrocytaire.

	Immunisation anti-érythrocytaire	Allo-immunisation	Auto-immunisation
<i>Nombre de patients</i>	27 (32,14 %)	14 (16,66 %)	16 (19,04 %)
<i>Spécificité des anticorps</i>		17 allo-anticorps CGRS : 1 anti-RH1, 1 anti-RH2, 3 anti-RH3, 1 anti-RH4, 3 anti-KEL1, anti-RH3 + anti-MNSI, anti-RH3 + anti KEL1 CGR P : anti-LE1, anti-FY1, anti-JK2	TCD type Ig G : 15 cas TCD type IgG + C' : 1 cas
<i>Nombre de syndromes drépanocytaires majeurs</i>	14 (25,92 %) NS	9 (16,66 %) NS	6 (11,11 %) NS
<i>Nombre de thalassémies</i>	13 (43,33 %)	5 (16,66 %)	10 (33,33 %)
<i>Sexe</i>	NS	NS	NS
Masculin	18 (35,29 %)	8 (15,68 %)	12 (23,52 %)
Féminin	9 (27,27 %)	6 (18,18 %)	4 (12,12 %)

CGRS : concentrés de globules rouges standard ; CGRP : concentrés de globules rouges phénotypés ; TCD : tests de Coombs directs.

Tableau 4

Immunsation anti-érythrocytaire et consommation de concentrés de globules rouges.

Patients (n = 84)			Syndromes drépanocytaires majeurs (n = 54)			Thalassémies (n = 30)		
Allo-immunsation (n = 14)	Auto-immunsation (n = 16)	Non immunisés (n = 57)	Allo-immunsation (n = 9)	Auto-immunsation (n = 6)	Non immunisés (n = 40)	Allo-immunsation (n = 5)	Auto-immunsation (n = 10)	Non immunisés (n = 17)
<i>Concentrés de globules rouges transfusés avant allo-immunsation</i>								
Total	231		140			91		
Moyenne	16,5		15,55			18,2		
Extrêmes	1-41		1-41			1-33		
<i>Concentrés de globules rouges transfusés avant auto-immunsation</i>								
Total	1472			172			1300	
Moyenne	92			28,66			130	
Extrêmes	2-220			2-77			6-220	
<i>Concentrés de globules rouges transfusés chez les non immunisés</i>								
Total		716			356			360
Moyenne		12,57			8,9			21,17
Extrêmes		1-132			1-45			1-132

La terapia trasfusionale nella talassemia

IMMUNOHEMATOLOGY

Clinically significant red blood cell antibodies in chronically transfused patients: a survey of Chinese thalassemia major patients and literature review

C.K. Cheng, C.K. Lee, and C.K. Lin

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CHENG ET AL.

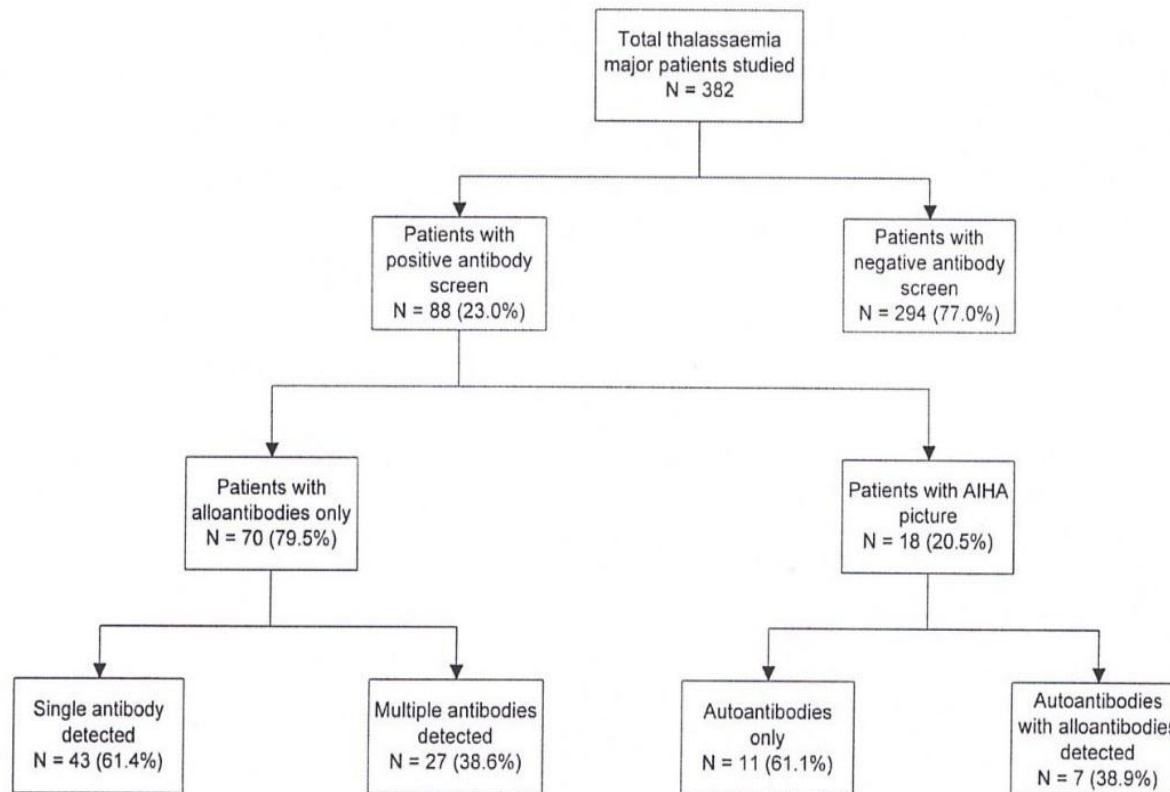


Fig. 1. Summary of the antibodies detected in the cohort of thalassemia major patients. Numbers in parentheses represent the percentages of total antibodies.

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TABLE 1. Specificities and frequencies of the 107 clinically significant alloantibodies in the 88 thalassemia major patients with positive antibody screen

Antibody specificity	Number	Frequency (%)
Rh system		
Anti-C	1	0.9
Anti-E	42	39.3
Anti-c	14	13.1
Anti-e	1	0.9
Anti-f	1	0.9
MNS system		
Anti-S	2	1.9
Anti-Mi ^a /Mur	33	30.85
Kell system		
Anti-K	1	0.9
Duffy system		
Anti-Fy ^b	2	1.9
Kidd system		
Anti-Jk ^a	7	6.55
Anti-Jk ^b	1	0.9
Diego system		
Anti-Di ^a	2	1.9

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TABLE 2. Specificities and frequencies of the seven clinically insignificant antibodies in the 88 thalassemia major patients with positive antibody screen

Antibody specificity	Number	Frequency (%)
Lewis system		
Anti-Le ^a	3	42.8
Anti-Le ^b	1	14.3
P blood group system		
Anti-P ₁	1	14.3
HLA system		
Anti-HLA	1	14.3
Anti-Bg	1	14.3

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TABLE 3. Summary of the 18 thalassemia major patients with warm autoantibodies

	Number	Frequency (%)
Autoantibodies only		
Male	7	38.9
Female	4	22.2
Autoantibodies with alloantibodies		
Male	4	22.2
Female	3	16.7

La terapia trasfusionale nella talassemia

ORIGINAL ARTICLE

Alloimmunisation in thalassaemics: a comparison between recipients of usual matched and partial better matched blood. An evaluation at a tertiary care centre in India

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La terapia trasfusionale nella talassemia

Pujani M et al

Table I - Clinical characteristics of the thalassaemics in the three groups.

	Group 1 (n=211)	Group 2 (n=46)	Group 3 (n=227)
Age			
<5 years	41 (19.4%)	45 (97.8%)	47 (20.7%)
6-10 years	82 (38.9%)	1 (2.2%)	88 (38.77%)
11-15 years	60 (28.4%)	-	51 (22.47%)
>15 years	28 (13.3%)	-	41 (18.06%)
Sex			
Males	128 (60.7%)	30 (65.21%)	139 (61.23%)
Females	83 (39.3%)	16 (34.79%)	88 (38.77%)
Diagnosis			
β thalassaemia major	207 (98%)	43 (93.48%)	220 (96.91%)
HbE- β thalassaemia	2 (0.95%)	3 (6.52%)	5 (2.2%)
Sickle- β thalassaemia	1 (0.5%)	-	1 (0.44%)
HbD- β thalassaemia	1 (0.5%)	-	1 (0.44%)
Splenectomy	Alloantibody development		
Yes	1/8 (12.5%)	0/0	1/16 (6.25%)
No	7/203 (3.5%)	-	7/211 (3.31%)
Age at start of transfusion	Alloantibody development		
≤ 1 year	4/64 (6.25%)	- (0/42)	4/122 (3.27%)
>1 year	4/147 (2.72%)	- (0/4)	4/105 (3.8%)

La terapia trasfusionale nella talassemia

Table II - Alloimmunisation rates and specificities of antibodies in all the groups.

	Alloimmunisation rate	Total n. of alloantibodies	Rh					Kell		Kidd
			D	C	C	Cw	E	K	Kp ^a	Jk ^a
Group 1 (UM)	3.79% (8/211)	10	1	-	1	1	3	2	1	1
Group 2 (PBM)	-	-	-	-	-	-	-	-	-	-
Group 3 (UM → PBM)	8	9	2	-	1	1	4	-	1	-

Blood Transfus 2014; 12 Suppl 1: s100-4 DOI 10.2450/2012.0154-12

La terapia trasfusionale nella talassemia

VoxSanguinis

The International Journal of Transfusion Medicine

ISBT International Society
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Vox Sanguinis (2014) 106, 197–208

REVIEW

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DOI: 10.1111/vox.12086

Red blood cell alloimmunization in sickle cell disease and in thalassaemia: current status, future perspectives and potential role of molecular typing

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La terapia trasfusionale nella talassemia

Isoimmunizzazione eritrocitaria prevenzione

*determinazione del fenotipo eritrocitario
all'esordio della malattia*

*terapia trasfusionale mediante concentrati eritrocitari
compatibili nell'ambito dei sistemi ABO , Rh e Kell
filtrati*

La terapia trasfusionale nella talassemia

Marker	Rischio di Infezioni Trasfusionali		
	HBV	HCV	HIV
	<i>Rischio per 1 milione di donazioni</i>		
Italia	69.1	16.7	1.9
Spagna	13.5	6.7	1.9
Francia	2.1	1.2	0.7

Gonzalez M et al,

Residual risk of transfusion-transmitted human immunodeficiency virus, hepatitis C virus, and hepatitis B virus infections in Italy, Transfusion, 2005



Epidemiology of HBV,HCV,HIV infections in patients with beta-thalassemia



COUNTRY	PATIENTS number	HBV n. pos	HCV n. pos	HIV n. pos
Turkey *	399	3 0.75%	18 4.5%	0
Iran**	732	11 1.5%	141 19.3%	0
India***	20	5%	5%	0

*Ocak, Arch Med Res, 2006 **Mirmomen S, Arch Iran Med, 2006

*** Chakrabarti S, Indian J Public Health,2006

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Arch Virol (2011) 156:1111–1115

DOI 10.1007/s00705-011-0950-y

ORIGINAL ARTICLE

Post-transfusion-transmitted hepatitis C virus infection: a study on thalassemia and hemodialysis patients in southeastern Iran

Gholamhossein Hassanshahi · Mohammad Kazemi Arababadi · Shokrollah Assar ·

Hamid Hakimi · Mozghan Noroozi karimabad · Mehdi Abedinzadeh ·

Houshang Rafatpanah · Reza Derakhshan

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1113

Table 2 The average serum levels of liver enzymes of HCV-infected thalassemia patients

Enzyme	Normal	Above normal	Statistical analysis
ALK	12 (44.5%)	15 (55.5%)	$P < 0.02$
SGOT	0 (0%)	27 (100%)	$P < 0.001$
SGPT	8 (29.7%)	19 (70.3%)	$P < 0.001$
DB	15 (55.5%)	12 (44.5%)	$P < 0.02$
TB	12 (44.5%)	15 (55.5%)	$P < 0.02$

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Table 1 The average age, sex and duration of transfusion of HCV-infected and uninfected thalassemia patients

	HCV positive N (%)	HCV negative N (%)	Statistical comparison
Average age	81 (44.75%) 14.50 ± 6.07	100 (55.25%) 10.13 ± 5.18	P < 0.05
Duration of transfusion (months)	161.25 ± 70	110.31 ± 61.4	P < 0.05
Sex	40 (49.39 %) male	41 (50.62 %) male	P > 0.05
	41 (41%) female	59 (59%) female	P > 0.05

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1114

G. Hassanshahi et al.

Table 3 The average age, sex and duration of hemodialysis of HCV-infected and uninfected hemodialysis patients

	HCV positive N %	HCV negative N %	Statistical comparison
N (%)	64 (31.5%)	139 (68.5%)	
Average age	51.16 ± 18.52	50.50 ± 20.44	P > 0.05
Duration of hemodialysis (months)	16.56 ± 9.89	15.57 ± 9.23	P < 0.05
Sex	33 (38.8%) male	52 (61.2%) male	P > 0.05
	31 (26.3%) female	87 (73.7%) female	P > 0.05

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EUROPEAN JOURNAL OF
Haematology

European Journal of Haematology 90 (501-507)

ORIGINAL ARTICLE

Natural history of hepatitis C in thalassemia major: a long-term prospective study

Maria E. Lai¹, Raffaella Origa², Fabrice Danjou², Gian B. Leoni², Stefania Vacquer¹, Franco Anni²,
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¹Ospedale Regionale per le Microcitemie, ASL8, Cagliari; ²Clinica Pediatrica 2a, Cagliari, Italy; ³Laboratory of Infectious Diseases, National Institute of Allergy and Infectious Diseases, National Institutes of Health, Bethesda, MD, USA

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Hepatitis C in thalassemia major

Eliana *et al.*

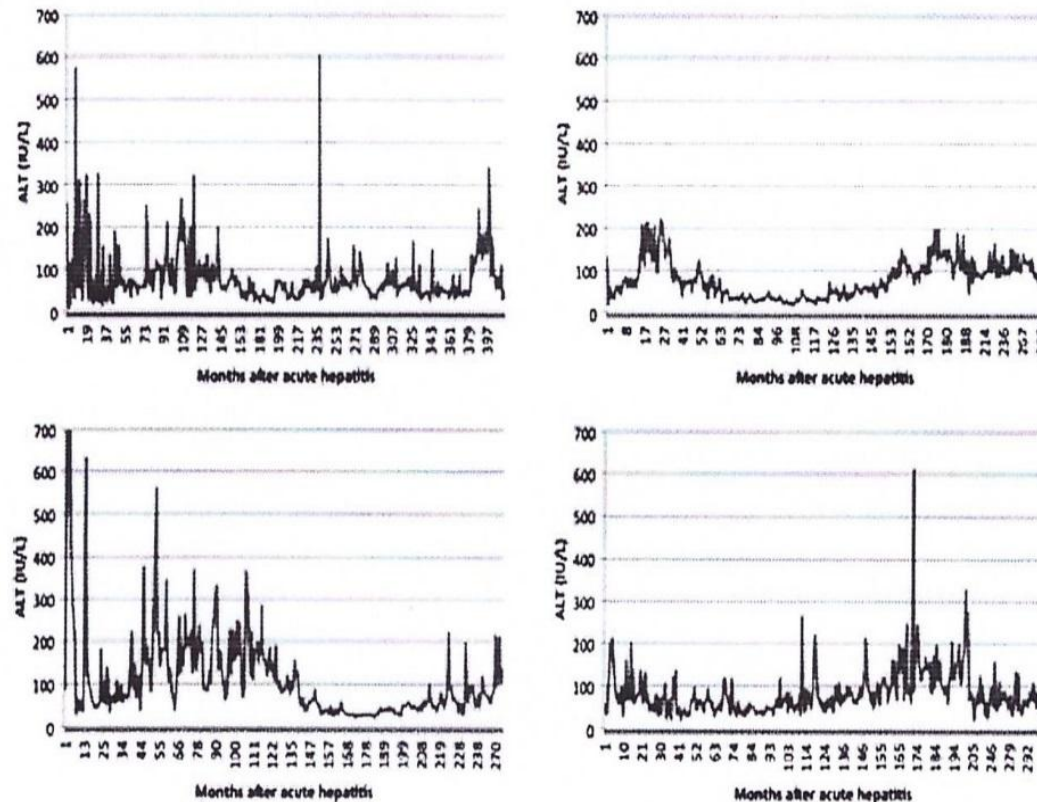
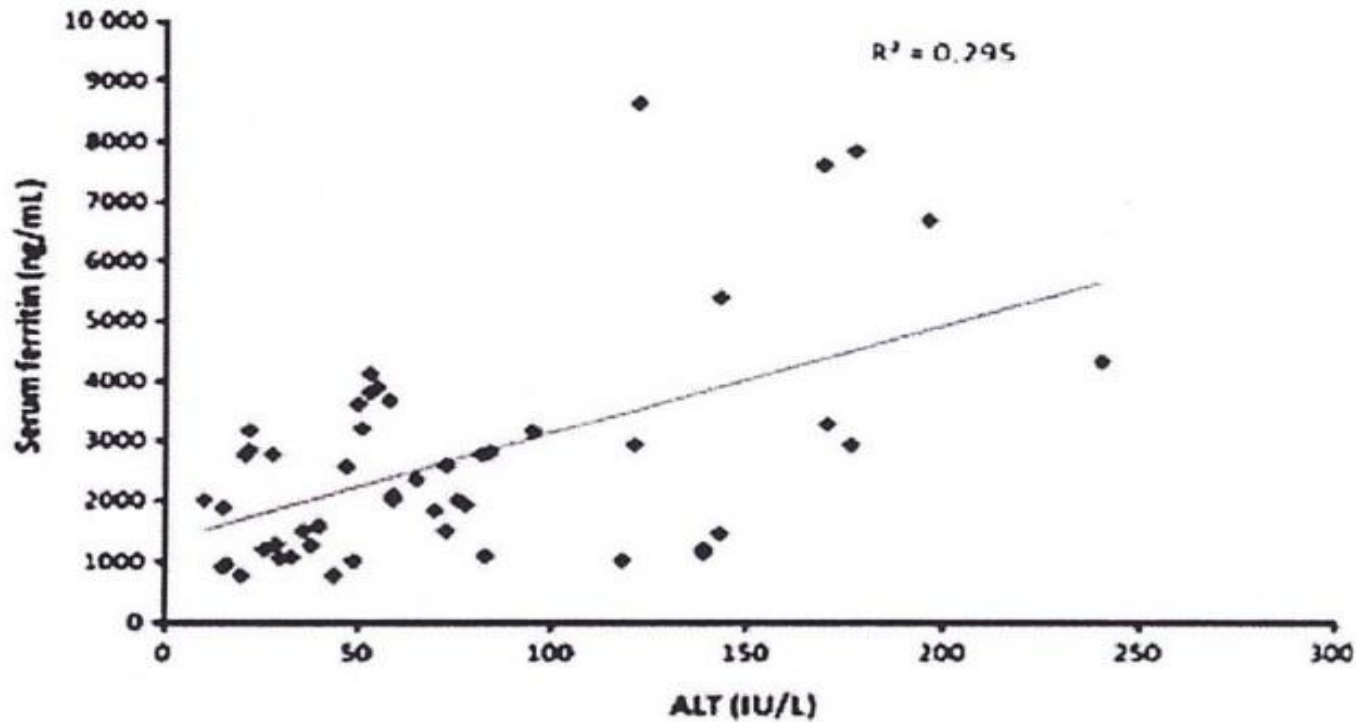


Figure 2 ALT follow-up in four patients with thalassemia and chronic hepatitis C since the time of viral infection. Each graph represents a single patient; ALT values were measured monthly.

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La terapia trasfusionale nella talassemia

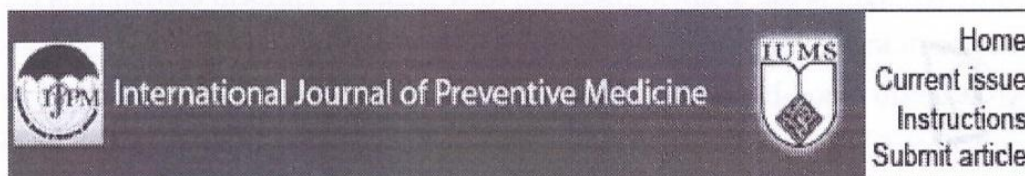
Eliana *et al.*

Hepatitis C in thalassemia major

Table 2 Liver biopsy findings according to HCV RNA status

Fibrosis score (Desmet <i>et al.</i>) (25)	Number and percentage of patients	Mean time interval between acute hepatitis and liver biopsy (years)	Number of cases upon iron score (Sciot <i>et al.</i>) (26)			
			I	II	III	IV
I						
HCV RNA positive	6 (31.6%)	17.3 ± 3	1	1	3	1
HCV RNA negative	7 (53.8%)	14.1 ± 6.1	0	5	0	2
II						
HCV RNA positive	8 (42.1%)	14.99 ± 3.4	0	0	4	2
HCV RNA negative	3 (23.1%)	11.74 ± 1.95	0	0	1	2
III						
HCV RNA positive	4 (21.0%)	15.84 ± 4.1	0	0	2	2
HCV RNA negative	3 (23.1%)	11.35 ± 5.29	0	0	0	3
IV						
HCV RNA positive	1 (5.3%)	17	0	0	0	1
HCV RNA negative	0		0	0	0	0

La terapia trasfusionale nella talassemia



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Prevalence of Anti HCV Infection in Patients with Beta-Thalassemia in Isfahan-Iran

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La terapia trasfusionale nella talassemia

Prevalence of Anti HCV Infection in Patients with Beta-Thalassemia in Isfahan-Iran

Pagina 6 di 8

Table 1

Variable	No. of Subjects		HCV ⁺		P-value
	N	%	N	%	
Sex					0.4
Male	270	58.3	24	8.9	
Female	193	41.7	13	6.7	
Residence					0.11
Urban	365	78.8	33	9	
Rural	98	21.2	4	4.1	
employment status					0.82
Employment	57	12.3	5	8.8	
unemployment	405	87.7	32	7.9	
Marital status					1
Married	38	8.2	3	7.9	
Single	425	91.8	34	8	
Education					0.006
Illiterate	13	3.1	0	0	
Elementary	83	19.7	1	1.2	
Guidance	85	20.1	9	10.6	
High school	177	41.9	16	9	
University	64	15.2	11	17.2	
Age (Mean $\pm$ SD)	17.46 $\pm$ 8.32		23.73 $\pm$ 7.33		<0.001

Univariate association between demographic characteristics and HCV seropositivity

La terapia trasfusionale nella talassemia

Prevalence of Anti HCV Infection in Patients with Beta-Thalassemia in Isfahan-Iran

Pagina 7 di 8

Table 2

Variable	No. of Subjects		HCV ⁺		P-value
	N	%	N	%	
History of tattooing					0.25
Yes	12	2.6	2	16.7	
No	451	97.4	35	35	
History of cupping					0.22
Yes	3	0.6	1	33.3	
No	460	99.4	36	7.8	
History of ear piercing					0.43
Yes	191	41.3	13	6.8	
No	272	58.7	24	8.8	
History of surgery					0.01
Yes	149	32.3	19	12.8	
No	312	67.7	18	5.8	
History of dental procedure					0.005
Yes	321	69.5	33	10.3	
No	141	30.5	4	2.8	
History of illegal sex					0.38
Yes	17	3.7	0	0	
No	442	96.3	37	8.4	
History of prison					1
Yes	1	0.2	0	0	
No	458	99.8	37	8.1	
Number of units transfused ^a (Mean ± SD)	2.03 ± 1.39		2.78 ± 2.15		<0.001
Number of transfusion ^a (Mean ± SD)	1.43 ± 1.82		1.51 ± 0.81		0.047
Duration of transfusion ^b (Mean ± SD)	16.02 ± 7.75		21.84 ± 5.77		<0.001

a; per month, b; years

Univariate association between potential risk factors and HCV seropositivity

La terapia trasfusionale nella talassemia

Complicanze infettive prevenzione

*donatore volontario periodico
anamnesi accurata del donatore*

*ricerca dell'infezione virale mediante biologia molecolare
inattivazione virale*



First Mediterranean Meeting on Blood Transfusion in Thalassemia, 1984



Centro Trasfusionale Universitario

Intervallo fra i diversi episodi trasfusionali in talassemici

Paziente	Regime	
	Ipertrasfusione (giorni)	Neociti (giorni)
CF	24	28
ME	27	33
VC	22	26
MS	21	31
PP	19	21
PM	21	23
RA	22	27
PC	27	28
DS	22	28

La terapia trasfusionale nella talassemia

Indian J. Hematol. Blood Transfus 24(4):173–177

ORIGINAL ARTICLE

Transfusion of neocytes concentrate/pooled neocytes in β -thalassemic patients

Dharmesh Chandra Sharma · Sunita Rai · Navneet Agarwal · Satya Sao · Ajay Gaur · Rahul Sapra

La terapia trasfusionale nella talassemia

Indian J. Hematol. Blood Transfus 24(4):173–177

175

Table 1 Comparison of control and study group

Groups	Transfusion interval in days	No. of transfusion	Serum ferritin fall (%)	Serum ferritin Increase (%)
Control	22–30 (Avg. 26.15)	14	–	9.6
Study	32–54 (Avg. 45.86)	08	20.1	–

Avg. = Average

La terapia trasfusionale nella talassemia

Table 2 Transfusion of conventional packed RBC units Group-I: control group

Sr. No.	Age in Years	Avg. transfusion interval in days	S. Ferritin level before study (ng/ml)	S. Ferritin level after study (ng/ml)	Increase in Ferritin level (%)	No. of transfusions	Total volume transfused in mL
1.	3½	24.33	2200	2420	9.1	15	2200
2.	5	26.07	3900	4280	8.08	14	2800
3.	7½	28.07	4012	4423	9.3	13	3120
4.	7	28.07	3982	4490	11.4	13	2660
5.	6	24.33	2850	3135	9.1	15	3750
6.	4	26.07	3100	3421	11.03	14	2530
Avg.		26.15	3340.66	3694.83	9.6	14	2843

Avg. =Average; S. = Serum.

La terapia trasfusionale nella talassemia

Table 3 Transfusion of neocyte/pooled neocytes units Group-II: study group

Sr. No.	Age in years	Avg. transfusion interval in days	S. Ferritin level before study (ng/ml)	S. Ferritin level after study (ng/ml)	Decrease in Ferritin level (%)	No. of transfusions	Total volume transfused in ml.
1.	4½	45.62	2920	2328	20.27	8	1600
2.	6	52.14	4100	3275	20.12	7	1610
3.	8	45.62	4104	3278	20.12	8	2160
4.	3	40.55	2210	1760	20.36	9	1440
5.	5	45.62	3425	2745	19.85	8	1760
6.	4	45.62	3102	2482	19.99	8	1520
Avg.	45.86	3310.16	2644.66	20.11	8	1681	

Avg. =Average; S. = Serum.

Conclusioni

*La prevenzione delle principali complicanze trasfusionali
è possibile e consente un ulteriore aumento della qualità di vita*



Isoimmunizzazione in neonati

Author	Year	Rbc Ab / patients	Rbc Ab /transfusions
<i>Strauss</i>	<i>2000</i>	<i>0/30</i>	<i>0/139</i>
<i>Floss</i>	<i>1986</i>	<i>0/53</i>	<i>0/683</i>
<i>Ludvigsen</i>	<i>1987</i>	<i>0/90</i>	<i>0/1269</i>
<i>Pass</i>	<i>1976</i>	<i>0/94</i>	<i>nd</i>
<i>Rawls</i>	<i>1984</i>	<i>0/126</i>	<i>nd</i>

La terapia trasfusionale nella talassemia

Table 3. Logistic regression analysis

	B	Standard error (SE)	Wald- chi-squared	P-value
DCT	1.7	1.063	2.84	0.09
Hb	-0.314	0.31	1.01	0.31
Duration of transfusion	-0.092	0.061	2.262	0.13
Age	0.082	0.093	0.77	0.38
Blood group			4.20	0.75
A positive	18.186	17284.15	0.00	0.99
AB negative	41.941	43751.76	0.00	0.99
AB positive	-0.507	19741.56	0.00	1.00
B negative	20.667	17284.15	0.00	0.99
B positive	17.911	17284.15	0.00	0.99
O -positive	19.030	17284.15	0.00	0.99
O negative	22.009	17284.15	0.00	0.99
Spleen			0.12	0.94
Enlarged spleen	0.157	0.97	0.02	0.87
Splenectomy	-0.436	1.67	0.06	0.79
Constant	-37.622	18315.16	0.00	0.99

Hb, haemoglobin.

La terapia trasfusionale nella talassemia

Table 2. Frequency of RBC antigens in the local donor population and RBC phenotype available on 20 Asian thalassemia patients

RBC antigen	All local donors (n = 802)	White donors (n = 352)	Asian donors (n = 103)†	Asian donors (n = 435)†	Asian patients (n = 20)	Average antigen frequency (Asians)	P
s*	93.1	91.7	90.5	99.5	100	97.86	—
Fya*	77.1	60.2	89.5	98.2	95	96.48	—
C*	51.7	50.5	84.4	95.7	95	93.59	—
E*	95.5	95.4	88.5	93.7	100	92.97	—
M	NA	NA	NA	83	75	82.65	—
Jkb*	57.1	70.4	67	81	85	78.56	—
N	NA	NA	NA	72	53.8	71.20	—
Jka*	53.7	69.3	68	68	75	68.25	—
Leb	NA	NA	NA	64	66.6	64.11	—
C*	80.4	77.8	43.7	53.8	35	51.26	< .0001
E*	21.4	19	32.0	43	25	40.32	—
P1*	NA	NA	NA	40	33.3	39.71	—
Lea	NA	NA	NA	20	40	20.88	—
Fyb*	63	78.7	16.8	18	20	17.85	< .0001
S*	40.5	51.4	15.6	12.7	20	13.50	.001
K*	—‡	9‡	0	0	1.0	0.3	< .0001

All local donors include mostly white (85%) and other ethnicities (15%). Asian donor phenotypes were collected during 2 separate time periods. Significant antigen frequency differences between white and Asian RBCs are on the rows with *P* values given. *P* was calculated for the antigens appearing in a higher frequency among the white (and total) donor population compared to the average frequency found among Asian patients. NA indicates not available.

*Indicates clinically significant antibodies (ie, can cause a hemolytic transfusion reaction or hemolytic disease of the newborn).

†Donors' phenotypes were collected in 2 separate time periods.

‡Frequency of Kell (K) antigen was not always examined. Therefore the known frequency among white people was used.

Alloimmunization and erythrocyte autoimmunization in transfusion
dependent thalassemia patients of predominantly Asian descent
ST Singer et al, Blood, 2000

La terapia trasfusionale nella talassemia

Table 1 Genetics of 33 blood group systems [6,7,12]

System	ISBT symbol	ISBT number	Chromosome location	N. antigens	Gene names	N. alleles
ABO	ABO	001	9q34.2	4	<i>ABO</i>	306
MNS	MNS	002	4q31.21	46	<i>GYPA, GYPB, GYPE</i>	47
P1PK	P1PK	003	22q13.2	3	<i>A4GALT</i>	48
Rh	RH	004	1p36.11	52	<i>RHD, RHCE</i>	361
Lutheran	LU	005	19q13.32	20	<i>LU, BCAM</i>	20
Kell	KEL	006	7q34	34	<i>KEL</i>	58
Lewis	LE	007	19p13.3	6	<i>FUT3</i>	74
Duffy	FY	008	1q23.2	5	<i>FY, DARC</i>	11
Kidd	JK	009	18q12.3	3	<i>JK, SLC14A1</i>	25
Diego	DI	010	17q21.31	22	<i>DI, SLC4A1</i>	91
Yt	YT	011	7q22.1	2	<i>YT, ACHE</i>	4
Xg	XG	012	Xp22.33	2	<i>XG</i>	2
Scianna	SC	013	1p34.2	7	<i>SC, ERMAP</i>	9
Dombrock	DO	014	12p12.3	8	<i>DO, ART4</i>	22
Colton	CO	015	7p14.3	4	<i>CO, AQP1</i>	12
Landsteiner–Wiener	LW	016	19p13.2	3	<i>LW, ICAM4</i>	3
Chido/Rodgers	CH/RG	017	6p21.32	9	<i>CH/RG, C4A, C4B</i>	7
Hh	H	018	19q13.33	1	<i>FUT1</i>	104
Kx	XK	019	Xp21.1	1	<i>XK</i>	35
Gerbich	GE	020	2q14.3	11	<i>GE, GYPC</i>	10
Cromer	CROM	021	1q32.2	17	<i>CROM, CD55</i>	15
Knops	KN	022	1q32.2	9	<i>KN, CR1</i>	32
Indian	IN	023	11p13	4	<i>IN, CD44</i>	4
Ok	OK	024	19p13.3	3	<i>OK, BSG</i>	5
Raph	RAPH	025	11p15.5	1	<i>RAPH, CD151</i>	4
John Milton Hagen	JMH	026	15q24.1	6	<i>JMH, SEMA7A</i>	12
I	I	027	6p24.2	1	<i>GCNT2</i>	13
Globoside	GLOB	028	3q26.1	1	<i>B3GALNT1</i>	9
Gill	GIL	029	9p13.3	1	<i>GIL, AQP3</i>	8
Rh-associated glycoprotein	RHAG	030	6p12.3	4	<i>RHAG</i>	23
Forssman	FORS	031	9q34.2	1	<i>GBGT1</i>	5
JR	JR	032	4q22.1	1	<i>JR, ABCG2</i>	19
LAN	LAN	033	2q36	1	<i>LAN, ABCB6</i>	15

200 A. Matteucci & L. Pierelli

Table 2 Clinical applications of DNA-based assays for blood group typing

Setting	Applications	Subject
Transfusion Practice	Blood typing in recently transfusion therapy (mixed field agglutination)	Patient
	Blood typing in a positive direct antiglobulin test (DAT)	Patient
	Distinction between auto and alloantibodies	Patient
	Investigation of serological ABO discrepancies	Patient and donor
	Determination and/or confirmation of D status	Patient and donor
	Confirming rare or weakly expressed RBC antigens	Patient and donor
	Antigenic monitoring in allogeneic stem cell transplant	Patient and donor
	Typing for antigens for which there are no commercial reagents	Patient and donor
	Extended antigenic matching in haemoglobinopathies	Patient and donor
	High-throughput screening for rare or uncommon antigen inventory	Donor
Prenatal Practice	Manufacturing of RBC antibody identification panels	Donor
	Definition of D-variants for immunoprophylaxis administration	Mother
	RHD-Zygosity testing	Father
	Antigenic typing by amniocentesis	Foetus
	Non-invasive RHD genotyping from the maternal plasma	Foetus

La terapia trasfusionale nella talassemia

Table 3 Clinical significance of some alloantibodies to RBC antigens
(from Reid and Lomas-Francis modified)[12]

Usually clinically significant	Sometimes clinically significant	Clinically insignificant if no reactive at 37 °C	Generally clinically insignificant
A and B	Colton	A ₁	Chido/Rodgers
Diego	Cromer	H	JMH
Duffy	Dombrock	Le ^a	HLA/Bg
H in O _h (Bombay)	Gerbich	Lutheran	Knops
Kell	Jr ^a	M, N	Le ^b
Kidd	Landsteiner –Wiener	P ₁	Xg ^a
P, PP ₁ P ^k	Scianna		
Rh	Yt ^a		
S, s, U	Lan		
Vel			

La terapia trasfusionale nella talassemia

Table 3

Variable	OR	95% CI
History of tattooing		
Yes	1.5	0.24-9.32
No	1	-
History of cupping		
Yes	2.73	0.04-196.49
No	1	-
History of ear piercing		
Yes	2.21	0.02-329.83
No	1	-
History of surgery		
Yes	1.36	0.60-3.08
No	1	-
History of dental work		
Yes	1.87	0.60-5.91
No	1	-
History of illegal sex^a		
Yes	<0.001	-
No	1	-
History of prison^a		
Yes	0.54	-
No	1	-
Number of units transfused^b	1.16	0.97-1.38
Number of transfusion^b	1	0.77-1.23
Duration of transfusion^c	1	0.84-1.09

Variables shown in this table were controlled for age, sex, job, marriage and education. OR, odds ratio; CI, confidence interval

a; according to the no enough observation, model is not able to estimate parameters, b; per month, c; years

Multiple logistic regression of potential risk factors for HCV seropositivity

La terapia trasfusionale nella talassemia

Table 1 IL-28 genotypes and HCV clearance in 52 patients with thalassemia major infected during childhood

	Genotype	Anti-HCV+/HCV RNA–		Anti-HCV+/HCV RNA+	
		<i>n</i>	%	<i>n</i>	%
rs12980275	AA	15	71	7	27
	AG	5	24	14	54
	GG	1	5	5	19
rs8099917	GG	0	0	5	19
	GT	4	19	12	46
	TT	17	81	9	35
rs11881222	AA	16	76	8	29
	AG	5	24	15	54
	GG	0	0	5	18
rs12979860	CC	14	67	7	25
	CT	7	33	16	57
	TT	0	0	5	18

La terapia trasfusionale nella talassemia



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HEPATITIS MONTHLY

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Epidemiology of Occult Hepatitis B Infection Among Thalassemic, Hemophilia, and Hemodialysis Patients

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La terapia trasfusionale nella talassemia

Occult Hepatitis B Infection Among Thalassemic Patients

Arbabadi MK et al.

Table 1. The Prevalence of OBI and HBV Serological Markers Among Blood Recipient Patients

	Disease	Country	Region, City	Prevalence of OBI ^a , %	Prevalence, %		
					Anti HBc	Anti HBs	HBs Ag
Toyoda <i>et al.</i> (11) (2004)	Hemophilia	Japan	ND	51.2	86	62.8	0
Borhany <i>et al.</i> (12) (2011)	Hemophilia	Pakistan	Karachi	1.73	ND ^a	ND	ND
Windyga <i>et al.</i> (13) (2006)	Hemophilia	Polish	ND	0	69.5	49.5	7.8
Aghakhani <i>et al.</i> (18) (2010)	Hemodialysis	Iran	Tehran	3.11	6.2	77.5	2.8
Arbabadi <i>et al.</i> (2) (2009)	Hemodialysis	Iran	Kerman	0	33.3	40.7	0
Di Stefano <i>et al.</i> (20) (2009)	Hemodialysis	Italia	ND	26.6	72	31	ND
Abu-El-Makarem <i>et al.</i> (22) (2012)	Hemodialysis	Egypt	ND	4.13	20	ND	ND
Sav <i>et al.</i> (25) (2010)	Hemodialysis	Turkey	ND	16.9	ND	ND	ND
Kanbay <i>et al.</i> (24) (2006)	Hemodialysis	Turkey	ND	15.2	ND	ND	ND
Goral <i>et al.</i> (28) (2006)	Hemodialysis	Turkey	ND	0	ND	ND	0
Siagris <i>et al.</i> & Mina <i>et al.</i> (10, 29) (2010)	Hemodialysis	Greece	ND	0.9-20.4	47.8	66.9	5.5
Motta <i>et al.</i> (31) (2010)	Hemodialysis	Brasilia	ND	15	3	27	0
Gabrerizo <i>et al.</i> (30) (1997)	Hemodialysis	Italy	ND	58	ND	ND	ND
Gwak <i>et al.</i> (32) (2008)	Hemodialysis	South-Korea	ND	0	67.4	69.8	4.8
Jain <i>et al.</i> (23) (2008)	Hemodialysis	China	ND	5	5	ND	11
Singh <i>et al.</i> (17) (2003)	Thalassemia	India	ND	31.4	20	75.7	5.7
Arbabadi <i>et al.</i> (14) (2008)	Thalassemia	Iran	Kerman	0	33	40.7	0

^a Abbreviations: ND, not determinant; OBI, occult hepatitis B infection