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AHMEDABAD MEDICAL ASSOCIATION

(Branch of Indian Medical Association)

ESTD : 1902

AHMEDABAD MEDICO NEWS

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DR. DILIP B. GADHAVI
President - AMA

DR. GARGI PATEL
Hon. Secretary - AMA

Imm. Past President - DR. KIRITKUMAR C. GADHAVI

VOL. 16 | APRIL-2022 | ISSUE-12

HIGHLIGHTS

Programme

08-05-2022	AMA Sr. Citizen Club Programme	15-05-2022	Scientific Programme
15-05-2022	Webinar on International Family Day	29-05-2022	Scientific Programme

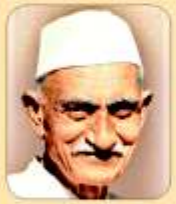
Article

Thalassemia - An Overview - **By Dr. Nidhi Bhatnagar**

ગુજરાત સ્થાપના દિવસ તા. ૦૧-૦૫-૧૯૬૦

નવાબ મહેંદી નવાઝ જંગ
પ્રથમ ગવર્નર - ગુજરાત

ડૉ. જીવરાજ મહેતા
પ્રથમ મુખ્યમંત્રી - ગુજરાત



રવિશંકર મહારાજ

ઈન્દુલાલ યાજ્ઞિક

'SECRET TO SUCCESS IS THE RESULT OF UNITY, HARD WORK AND PERSEVERANCE'

Message From President's & Hon. Secretary's Desk

Dear members,

When we think about the quote of Nobel laureate **Albert Einstein,** - "Only a life lived for others is a life worthwhile", always doctor will come first in majority of people's mind. But, when such virtuous persons are targeted by short sited & avaricious people of the society, it hurts a lot. There was an intense grief in every doctor's heart on unfortunate demise of Dr. Archana Sharma who was victimized by greedy people. The series of events arranged nationwide, which were not only to give her shrandhanjali but also to show our strong feeling of displeasure & dissatisfaction to entire nation. At Ahmedabad medical association also more than 500 doctors gathered on 2nd April. Though it was a peaceful shraddhanjali with keeping 2 minutes silence, candle march & by making a very long human chain, there was vexation in every mind. We have full sympathy with her family & give heartfelt condolences to them. But, if we see on the other side, if we remain united & in touch with our association, such regrettable event can be prevented.

We have continued organizing scientific events as well. Before above incidence happened, we have tried to address the issue of good patient-doctor relationship; a CME was arranged on 27th March 2022; Dr. Parimal Desai, Dr. Hitesh Patel & other speakers put their efforts to make everyone to understand this aspect of clinical practice also. Another CME was organized on 17th April 2022, in association with IMA HQ, "Respiratory update", covering the topics specifically on Allergy & UAD. This year we have planned a mega annual conference AMACON 2022. There will be wonderful scientific feast with renowned speakers. The conference will have many more features & attractions. We appeal the members to participate in the conference & make this event a grand success.

An entertainment programme was organized on 16th April 2022 evening; at AMA hall, Ashram road. It was tribute to Lataji in form of 16 selected songs in respect to life of woman. We are thankful to Dr. G. G. Oza sir, Dr. Gurudatt Thakkar, Dr. Urman Dhruv - Mrs. Titikshaben & Dr. Ashok Patel for making the event most enjoyable.

We wish everyone to keep yourself healthy in the extreme hot month of the year & be happy always.

Jay AMA

Jay IMA

Dr. Dilip B. Gadhavi
President
Ahmedabad Medical Association

Dr. Gargi Patel
Hon. Secretary
Ahmedabad Medical Association

CWC Meeting at Panchkula, Haryana on 16-17 April 2022

CWC Meeting at Panchkula, Haryana on 16-17 April 2022

Installation Ceremony of Physician Association of Ahmedabad & ANSHCON-2022 Dr. Dilip Gadhavi as a Chief Guest

Scientific Programme on 27-03-2022

Scientific Programme on 27-03-2022

Shraddhanjali & Candle -Light March on 02-04-2022

Shraddhanjali & Candle -Light March on 02-04-2022

Shraddhanjali & Candle -Light March on 02-04-2022

Entertainment Programme : Tribute to Lataji : 16-04-2022

Workshop by AMA with AMC on 22-03-2022

Scientific Programme on 17-04-2022

A.M.A. BULLETIN INDEX

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AHMEDABAD MEDICAL ASSOCIATION YEAR : 2021-2022

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9898047505

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9228233441

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DR. KAMLESH PATEL	DR. SNEHAL KALE	DR. DHIREN R. MEHTA	DR. VIPUL TURKHIA

NOTICE

A General Body Meeting of Ahmedabad Medical Association will be held on WEDNESDAY, 11th May, 2022 at 3.00 p.m. at our premises to transact the following agenda.

AGENDA

1. To read and confirm the minutes of the last meeting.
2. Any thing arising out of it.
3. Election, if required for the post of Vice President of Gujarat State Branch, I.M.A.
4. Any other business with the permission of the chair.

N.B. For the want of quorum the meeting will be adjourned and will be held after fifteen minutes.

1. Nominations are invited for the Post of Vice President (Ahmedabad Zone) of Gujarat State Branch, I.M.A. for the year 2022-2023.
2. Nomination forms will be available from the office from Monday 2nd May, 2022 on payment of Rs. 10/- (Rs. Ten only)
3. Nomination forms duly filled, proposed & seconded should reach Hon. Secretary on or before Saturday, 7th May, 2022, 3.00 P.M. at Association's office along with Rs. 250/- for the post of Vice President.
4. Last date of withdrawal of nomination is Monday, 9th May 2022 before 3.00 P.M.

Dr. Gargi M. Patel
Hon. Secreatry

Note :
1. He/She must be a life member of I.M.A.
2. He/She must have 7 years continuous membership of I.M.A.
3. He/She should have served G.S.B.I.M.A. as Working committee Member for at least 3 years.

AHMEDABAD MEDICAL ASSOCIATION
SENIOR CITIZEN CLUB SCIENTIFIC PROGRAMME

DATE : 08/05/2022, Sunday
TIME : 10.00 AM to 12.30 PM

Topic & Speakers

1. **મોટી ઉંમરે આરોગ્ય ની જાળવણી**
   Dr. Mahadev Desai
2. Measures to retain memory at old age
   Dr. Ruchir Divetia
3. Psycho-Social problems in Elderly
   Dr. Himanshu Desai
4. **વૃધ્ધ થવું એટલે સમૃધ્ધ થવું**
   શ્રી રમેશભાઈ તન્ના (આદરણીય લેખક અને પત્રકાર)

Chairpersons

Dr. K. R. Sanghvi Dr. K. C. Gadhavi

ઉપરોક્ત કાર્યક્રમમાં AMA ના સભ્યો, સીનીયર સીટીઝન ક્લબના સભ્યો તથા સૌ સભ્યોના spouse ભાગ લઈ શકે છે.

Registration fee Rs. 50/-
(કાર્યક્રમ અંગ્રેજી અને ગુજરાતીમાં છે)

Register at AMA office.

Programme will be followed by lunch.

AMA Senior Citizen Club, Co-ordinators

Dr. Abhay Dikshit Dr. K. R. Sanghavi
Dr. K. C. Gadhavi Dr. R. C. Shah

Dr. Dilip Gadhavi
President, AMA

Dr. Gargi Patel
Hon. Secretary, AMA

DR. SHANTILAL A SHAH SCIENTIFIC PROGRRAMME

WORLD ASTHMA DAY

DATE : 15/05/2022

TIME : 9.00 AM ONWARDS

VENUE : R M FOZDAR HALL, AMA HOUSE,
ASHRAM ROAD, AHMEDABAD

SPEAKER

Dr. TUSHAR PATEL,
PULMONOLOGIST

PROGRAMME CO ORDINATOR

Dr. ASHISH BHOJAK

REGISTER YOUR NAME AT AMA OFFICE BETWEEN
2.00 PM TO 6.00 PM
PHONE : 079 26588775

Dr. Dilip Gadhavi
President - AMA

Dr. Gargi Patel
Hon.Sec.- AMA

DR. K. L. VASA
SCIENTIFIC PROGRRAMME

Ahmedabad Medical Association
is organising a
Scientific Program
with reference to

WORLD NO TOBACCO DAY

DATE : 29/05/2022

TIME : 9.00 AM ONWARDS

VENUE : R M FOZDAR HALL, AMA HOUSE,
ASHRAM ROAD, AHMEDABAD

SPEAKERS

Dr. DEEPAK RAO
MS, MCh, Cancer Surgeons,
Pinaksh Hospital

Dr. DHRUV PATEL
MS, MCh, Cancer Surgeons,
Pinaksh Hospital

There will be a session on "Tobacco addiction - how to get rid of it" by well known psychiatrist of the city.

SPONSORED BY

PINAKSH CANCER HOSPITAL
3rd Floor, Avoin Building,
Beside Life Care Hospital,
Near Sardar Patel Statue,
Nathalal Colony, Navrangpura,
Ahmedabad - 380 014.

REGISTER YOUR NAME AT AMA OFFICE BETWEEN
2.00 PM TO 6.00 PM
PHONE : 079 26588775

Dr. Dilip Gadhavi	Dr. Gargi Patel
President - AMA	Hon.Sec.- AMA

WE WELCOME FOLLOWING NEW LIFE MEMBERS

12173	L	DR. VORA VISHVA JINAL
12174	L	DR. GAJJAR SHARVIL HETAVKUMAR
12175	L	DR. CHAREL SUJATABEN RAMABHAI
12176	L	DR. SHAH ADITYA SAUMIRBHAI

WEBINAR ON INTERNATIONAL FAMILY DAY

DATE : 15/05/2022

TIME : 4.00 PM ONWARDS

SPEAKERS

Dr. Pragnesh Vachharajani

Dr. Sucheta Shyamal Munshi

PROGRAMME CO ORDINATOR

Dr. KALPITA DAVE

Dr. Dilip Gadhavi
President - AMA

Dr. Gargi Patel
Hon.Sec.- AMA

SHRADDHANJALI

May his soul rest in eternal peace.

Padmashree Dr. Devendrabhai Patel

Date of Death : 05-04-2022

We send our sympathy & condolence to the bereaved family.

ANNOUNCEMENT
ANNUAL CONFERENCE OF
AHMEDABAD MEDICAL ASSOCIATION

AMACON – 2022

Dear members,

This year we have planned a Mega Conference AMACON–2022

Date	:	19-06-2022, Sunday
Time	:	9 am to 5 pm
Venue	:	J. B. Auditorium, Ahmedabad Management Association, ATIRA
Registration fees	:	Rs 1000/- per Delegate

It will be a whole day programme with excellent scientific feast. We will arrange simultaneous paper & poster presentation to project the research works by various doctors & PG students (IMA members). The delegates will get attractive gift, kits & many more. The more details will be shared shortly.

For registration, contact at office of Ahmedabad Medical Association in between 2 pm to 6 pm. Ph. : 079-26588775

Dr. Dilip Gadhavi
President - AMA

Dr. Gargi Patel
Hon.Sec.- AMA

FAMILY BENEFIT SCHEME
AHMEDABAD MEDICAL ASSOCIATION

ELIGIBILITY

- Life member of Ahmedabad Medical Association up to completion of 55 years of age.
- Spouse also eligble to become member
- For members above 40 years of age, 3 years of contineuous life membership of AMA is mandatory.

MEMBER BENEFITS

- Death Benefit: Nominee of member will get 500rs/member contribution in event of death of member.
- Permanent Disability Benefit: In the event of member become permanently disable due to some reason, he will get 500 Rs./member. Then he will cease to become member of the scheme

FEES SCHDULE :

AGE	A.F.C.	ADMISSION FEES	TOTAL
UP TO 35	3000	0	3000
36-40	3000	0	3000
41-45	3000	5500	8500
46-50	3000	6600	9600
51-55	3000	7700	10700

SPECIAL INCENTIVE TO MEMBERS BELOW 40 YEARS, NO JOINING FEES UP TO 30.6.2022.

UNIQUE FEATURES

- NON MEDICAL SPOUSE IS ALSO ELIGIBLE TO BECOME MEMBER
- PERMANENT DISABILITY BENEFIT FOR MEMBER
- LOW ADMISSION FEES

FOR MORE INFORMATION

- AHMEDABAD MEDICAL ASSOCIATION
  amagsbima@yahoo.co.in,
  91-079-26588775, 9726888775
  Monday to Saturday : 2pm-6pm

OBITUARY

May their soul rest in eternal peace.

DR. VINODBHAI M. SHAH
L-1147 M.B.B.S.
Date of Birth : 01-09-1942
Date of Death : 03-04-2022

DR. JAYENDRAPRASAD N. DAVE
L-0153 M.D., SKIN
Date of Birth : 17-08-1948
Date of Death : 24-03-2022

We send our sympathy & condolence to the bereaved family.

Shraddhanjali to Dr. Archana Sharma

An unlawful series of event at Rajasthan, stole the life of our colleague, Dr Archana Sharma. To give Shraddhanjali, the members of Ahmedabad Medical Association gathered in a very huge numbers. Following a very short notice, on 2nd April at 7.00 pm more than 500 doctors gathered at AMA, Ashram road; the gathered doctors gave her shraddhanjali by keeping 2 minutes silence. Afterwards, a peaceful candle march was done & a human chain was formed in a disciplined manner on the Ashram road. We gave a strong message to the society that such incidence shall not be tolerated. Our peaceful event was acknowledged by various print & electronic media.

Report of Entertainment programme on 16-03-2022

A unique musical programme - tribute to Bharat Ratna Lata Mangeshkar, "Nightingale of India"

"નારીનું જીવન સંગીત.... લતાજીના કંઠે" was organized on 16th April 2022 evening; at AMA hall, Ashram road. It was tribute to Lataji in form of 16 selected songs in respect to life of woman with wonderful coordinated work by Dr. G. G. Oza sir, Dr. Gurudatt Thakkar, Dr. Urman Dhruv & his wife Mrs. Titikshaben & Dr. Ashok Patel. High tea was served. More than 200 persons enjoyed the event.

Report of Scientific Programme on Vector Borne Diseases on 22/03/2022

Ahmedabad Medical Association with Ahmedabad Municipal Corporation organized a Workshop on Malaria Elimination and Dengue case Management on 22 th March, 2022 from 1 pm to 4 pm at Gujarat Sports club, Ahmedabad.

There were two speakers.

1) Dr. Nilay Suthar

   M.D (Med.)

   Professor of Medicine, NHL Mun. Med. College

   Subject : Diagnosis and treatment of Malaria with case management of Dengue as per guidelines.

2) Dr. G. C. SAHOO

   Ex. Reg. officer

   Govt. of India

   Subject : Malaria Elimination Strategy in India

It was very informative Scientific Programme and very well attended.

Report of Scientific programme on 27-03-2022

A scientific programme organized by AMA & college of General Practitioners on 27-3-2022 (Sunday), Time: 10.00 am to 12.30 pm at Hotel RE:GEN:TA INN, Near Ranip ST Bus Stand, Ahmedabad. Dr Parimal Desai talked on "Doctor-Patient Relationship"; Dr Hitesh Patel talked on "How to enjoy our Profession"; Mr Chandan Chaudhary from EKA Care talked about "Smart practice in today's era" & Mr Krushna Dave spoke on **"કવિતાપથી - એક ઉપચાર"**. All the presentations were wonderful. Dr Mehul Shelat coordinated the entire event nicely. Almost 50 doctors participated in the event.

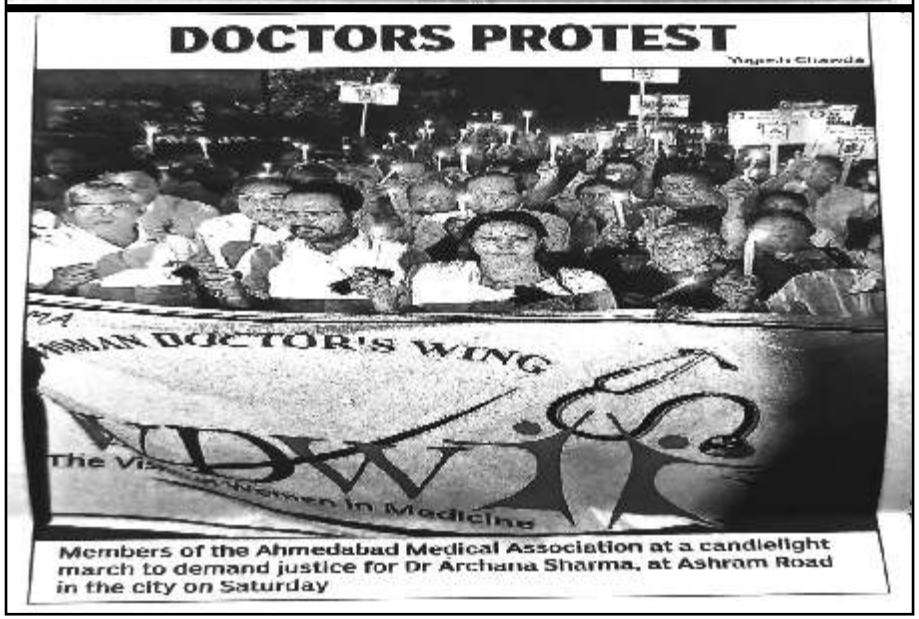
Report of Scientific programme on 17-04-2022

The Indian Medical Association HQ & Ahmedabad branch jointly organized a CME on Respiratory update in association with Sanofee India Pvt Ltd. On 17th April 2022, 9.00 am to 12.30 pm at AMA Hall, Ashram road, Ahmedabad. Dr Shweta Singh, ENT surgeon talked on "A to Z management of Allergic rhinitis – A comprehensive overview"; Dr Parthiv Mehta, Senior Pulmonologist & intensivist presented on "Multidisciplinary approach to UAD management- Zooming in the future". Both the presentations were very nice & very well elaborated. The lectures were followed by interactive question answer session which was moderated by Dr Tushar Patel, pulmonologist. Dr Urvesh Shah coordinated the CME. The CME was followed by delicious lunch. Total 70 members attended the CME. CME was accredited by Gujarat Medical Council.

રાજસ્થાનના મહિલા તબીબને ન્યાય અપાવવા કેન્ડલ માર્ચ

નવગુજરાત સમય

રાજસ્થાનના ગાયનેકોલેજિસ્ટ ડો. અર્ચના શર્માની આત્મહત્યાના મુદ્દે રાજ્યભરના તબીબોમાં ભારે રોષની લાગણી વ્યાપી છે. આ મહિલા તબીબને ન્યાય આપવાની માગણી સાથે ઇન્ડિયન મેડિકલ એસોસિએશન અને અમદાવાદ મેડિકલ એસોસિએશન દ્વારા આશ્રમ રોડ સ્ટેએમસે ખાતે બે મિનિટ મૌન પાળીને શ્રદ્ધાંજલિ અર્પણ કરી હતી અને ત્યારબાદ આશ્રમ રોડ પર મોટી સંખ્યામાં કેન્ડલ માર્ચ યોજી હતી. તસવીર: પંકજ શુક્લ



Members of the Ahmedabad Medical Association at a candlelight march to demand justice for Dr Archana Sharma, at Ashram Road in the city on Saturday

AHMEDABAD MEDICAL ASSOCIATION
HALL DONATION AND DEPOSIT CHART

Effect from 1-4-2016 Phone : 26588775 Office Time : 2 to 6 p.m.

Dr. R. M. Fozdar Hall (capacity 248 Seats)

	Others	members
Hall Deposit (Refundable)	Rs. 5,000-00	Rs. 5,000-00
Dr. R. M. Fozdar Hall (For 3 hours) Non A.C.	Rs. 4,500-00	Rs. 3,500-00
Extra Charges for 1 hour (Extra charges limit upto 3 hours)	Rs. 800-00	Rs. 700-00
A.C. Charges (For 3 hours)	Rs. 4,000-00	Rs. 3,500-00
(Extra Charges 1 hour) A.C.	Rs. 1,200-00	Rs. 1,000-00
Dr. R.M. Fozdar Hall Full Day (9 a.m. to 7 p.m.) Non A.C.	Rs. 11,000-00	Rs. 9,000-00
A.C. Full Day - Dr. R. M. Fozdar Hall	Rs. 11,000-00	Rs. 9,000-00

OPEN GROUND WITH Dr. R. M. Fozdar Hall

For Lunch / Dinner	Rs. 5,000-00	Rs. 4,000-00
For Refreshment	Rs. 1,200-00	Rs. 1000-00
Cleaning + Electric	Rs. 700-00	Rs. 600-00

JAGMOHAN PARIKH HALL 1ST FLOOR
(Capacity 100 Chairs)

Hall Deposit (Refundable)	Rs. 4,000-00	Rs. 4,000-00
J. P. Hall (For 3 hours) Non A.C.	Rs. 3,000-00	Rs. 2,500-00
Extra charges for 1 hour (Extra charges limit upto 3 hours)	Rs. 700-00	Rs. 600-00
A.C. Charges (For 3 hours)	Rs. 2,000-00	Rs. 1,500-00
(Extra charges 1 hours) A.C.	Rs. 600-00	Rs. 500-00
J. P. Hall Full day 9 a.m. to 7 p.m. (Non A.C.)	Rs. 7,000-00	Rs. 6,000-00
J. P. Hall A.C. Full Day	Rs. 6,500-00	Rs. 5,500-00

OPEN GROUND WITH DR. J. P. HALL

For Lunch / Dinner	Rs. 2,500-00	Rs. 2,000-00
For Refreshment	Rs. 700-00	Rs. 600-00
Cleaning + Electric	Rs. 500-00	Rs. 400-00

Dr. R. M. Fozdar Hall, J.P. Hall & Open Ground is not permitted for following purpose.

• Political programme Music programme (Professional) Marriage & Reception Event

Token Rates applicable for :

• GSB • IMA • S.S.S. • P.P.S. • N.S.S.S. Health Scheme Ladies Club

RATES FOR ADVERTISEMENT IN AMA BULLETIN

WITH EFFECT FROM 5-6-2013

PARTICULARS	AMA MEMBER	GROUP MEMBER	NON MEMBER CORPORATE HOSP.
Last Cover Page (Colour)	Rs. 10000	Rs. 11000	Rs. 12000
Inside Cover Page Total 4 in (Colour)	Rs. 8000	Rs. 9000	Rs. 10000
Center Page - the Rates are per page (Colour)	Rs. 8000	Rs. 9000	Rs. 10000
Full Page Colour	Rs. 5500	Rs. 7000	Rs. 9000
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SASHAKT KISHORI SASHAKT NARI SOCH BADALO KHUDKO BADLO DUNIYA BADLO

PROGRAM DETAILS

PROGRAM 1

DATE : 26 April, 2022, Tuesday

TIME : 10am to 12 noon

VENUE : Gulbai Tekra Ved Bhavan

Health Checkup Camp

DOCTOR : Dr. Praful Panagar, AMA

Ladies Club is Conducting free Health Checkup as Social Service

❀ ❀ ❀

PROGRAM 2

DATE : 29 April, 2022, Friday

TIME : 6.00pm to 7.00 pm.

VENUE : AMA House, Ashram Road.

YOGA TEACHER : Rachanaben Shah

FOOD : High Tea

PROGRAM 3

DATE : 06 May 2022, Friday

TIME : 6.30 pm onwards

VENUE : Dr. R M Fozdar Hall, AMA,

Ashram Road.

Entertainment Event Music Masti

Singing Karaoke & Dance

SPONSOR : Exide Finance

FOOD : Dinner - High Tea

Dr. Praful Panagar	Mrs. Khushboo Doshi
President - Ladies Club	Hon. Secretary - Ladies Club
M. 9327019168	M. 9429922071
Dr. Dilip B. Gadhavi	Dr. Gargi M. Patel
President - AMA	Hon. Secretary - AMA

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Thalassemia- An Overview

Introduction

The thalassemia are a group of congenital anemias that have in common deficient synthesis of one or more of the globin subunits of normal human haemoglobin. The primary defect is usually quantitative, consisting of the reduced or absent synthesis of normal globin chains. It is considered one of the most common genetic disorder worldwide. It occurs in a particularly high frequency in a broad belt extending from the Mediterranean basin through the Middle East, Indian subcontinent, Burma, Southeast Asia and islands of Pacific. About 3% of the world's population carry β-thalassemia genes. In Indians, frequencies between 3.5 % and 14.5% have been reported.

The thalassemias are genetically transmitted disorders. Normally an individual inherits two β-globin genes located one each on two chromosomes 11, and two alpha globin genes one each on two chromosome 16, from each parent i.e. normal adult hemoglobin is $\alpha_2\beta_2$. Depending upon whether the genetic defects or deletion lies in transmission of α or β globin chain gene, thalassemias are classified into α and β-thalassemias. They may occur as heterozygous (minor) or homozygous state (major). The former is generally asymptomatic while the later is severe congenital hemolytic anemia.

The alpha thalassemias are most commonly due to deletion of one or more of the alpha chain genes located on short arm of chromosome 16. In this disorder, there is defective synthesis of α-globin chains resulting in depressed production of hemoglobin that contains alpha chains i.e. HbA, HbA_2 and HbF. In β-thalassemia, there is decreased rate of β-chain synthesis resulting in reduced formation of HbA in the red cells. Most of β-thalassemias arise from different types of mutations of β-globin gene resulting from single base changes. Each of two main types of thalassemias may occur as heterozygous (minor) or homozygous state (major). The former is generally asymptomatic while the later is severe congenital hemolytic anemia. These subgroups have in common an imbalanced globin produced in excess is responsible for ineffective erythropoiesis and hemolysis.

Clinical Features

The clinical features of thalassemia major include features that are due to the disease itself and others that represent the consequences of the therapy. The early symptoms of the disease that is anemia appear usually in the first year of life. The infants fail to thrive and may have bouts of fever, diarrhoea and other gastrointestinal symptoms. The course of the disease in childhood depends almost entirely on whether the child is maintained on adequate transfusion program. Untransfused or poorly transfused patients with thalassemia develop typical bone abnormalities due

to the extremely increased erythropoiesis with consequent expansion of the bone marrow to 15 to 30 times normal. The skull is large and deformed by frontal and posterior bossing with the diploe increased in thickness to several times normal. Metatarsal, metacarpal, ribs and vertebral bones also expand as a consequence of increased erythropoiesis. Osteoporotic changes in bones due to bone marrow expansion, endocrine dysfunction and iron overload have been observed in thalassemia patients. Gall stones, thrombotic complications and pseudoxanthoma elasticum are also observed in these patients. Clinical manifestations of iron overload due to repeated blood transfusion therapy are common complications even in child who has received chelation therapy. The first indication of iron loading usually is the absence of the pubertal growth spurt and failure of the menarche. Over the succeeding years, a variety of endocrine disturbances may develop, particularly diabetes mellitus, hypogonadotrophic hypogonadism and growth hormone deficiency. Hypothyroidism and adrenal insufficiency also occur. Cardiac siderosis may cause an acute cardiac failure with arrhythmia or intractable cardiac failure. Liver parenchymal siderosis is also present from the very early stages of iron loading.

Laboratory Diagnosis

Laboratory findings at presentation are low haemoglobin with Mean Corpuscular Volume (MCV) typically 60 to 70 fl and

Mean Corpuscular Hemoglobin (MCH) 12 to 18 pg/cell. There is great variation in size and shape of erythrocyte in peripheral smear examination. High performance liquid chromatography findings are characterized by HbF levels ranging from 10 to 100%; HbA2 may be normal or increased up to 5 to 7% and remaining percentage is HbA. In the case of transfused patients, diagnosis can be made by globin chain synthesis analysis from peripheral reticulocytes or by β-globin gene analysis to identify various DNA mutations.

Current Management

Current disease management of β-thalassemia consists of prenatal diagnosis, transfusion therapy and allogeneic bone marrow transplantation (BMT), of which Allogeneic stem cell transplant remains the only option for a cure for β thalassemia major patients. The chance of a successful transplant is >90% in patients with good risk features while the outcome is still challenging for high risk patients. Because of the complications like graft-versus-host disease and transplant-related mortality the use of allogenic bone marrow is limited. The high cost is also not affordable by majority of the families with a thalassemia major child. Thus, prevention of the birth of an affected child is a feasible and realist option.

Regular blood transfusions, iron chelation in an attempt to prevent iron overload, judicious use of splenectomy in cases complicated by hypersplenism and a good standard of general

pediatric care are the treatment options available for the majority of thalassemic children in India. Children with thalassemia who are maintained at Hb level of 9.5 to 10 gm/dl constantly through hypertransfusion, grow and develop normally without skeletal abnormality. However there are advantages and disadvantages of hypertransfusion protocol. Iron overload is one of them. Maintaining a lower Hb level than this range without any deleterious effects on development with the added advantage of reducing the level of iron loading may be possible. Usually blood transfusions are given every 4 weeks on an outpatient basis. Red cell antigen phenotype matched leukocyte reduced packed red blood cells is always preferable for transfusion to reduced frequency of alloimmunization and transfusion reactions. Assessment of iron store by monitoring serum iron, serum ferritin and transferrin saturation and iron chelation therapy must be started within first 2 to 3 years of life to prevent iron loading toxicity.

Blood Transfusion Therapy

Blood Transfusion remains the mainstay therapy for a thalassemia major child. The decision to start transfusion depends upon Molecular diagnosis indicating severity of the disease, on the level of Hb and on the general conditions and satisfactory growth. The age of starting transfusion varies according to thalassemia genotype. Generally, the Hyper Transfusion Protocol is followed

where the Hb level is never allowed to fall below 9.5 to 10 gm/dl. The Pretransfusion Hb level should be 9 to 10 gm/dl and post transfusion Hb level should be 12 to 13 gm/dl.

The choice of Red cells should be Leukoreduced and Rh, Kell, Duffy and Kidd antigen matched. The red cell unit should be preferably less than 7 days old and haematocrit of unit should be 80% without additive solution or 60% with additive solution. The Transfusion Rate should be 5 to 6 ml/kg/hour and for a patient with cardiac failure, it should be 3 to 4 ml/kg/hour.

The Red cell requirement can be calculated as follows:

$(Hb^d - Hb^c)(3/\text{Hct of unit})(\text{Pt's wt. in Kg}) = \text{Milliliters of red cells to transfuse}$

[Hb^d - Desired Hb, Hb^c - Current Hb]

A Record of total amount of blood transfusions should be kept to calculate the iron intake and appropriate chelation therapy should be advised.

What does the future hold?

Increasing the haemoglobin F production by an increased globin chain production using different therapeutic agents e.g. 5-Azacytidine, hydroxyurea, cytarabine, vinblastine and butyrate and its analogue is promising. Due to inconsistent results and concerns about the potential for serious side effects from the long term use

these drugs, use of HbF inducers remains experimental and cannot be recommended outside of clinical trials.

As thalassemia is genetically derived disorder, genetic and cellular targets are potential approaches in management of disease. Currently the therapy is mainly directed at gene transfer into potential hematopoietic stem cells using retroviral vectors. Other approaches also are being taken, including attempts at the restoration of normal splicing in cases of splicing mutations and use of trans-splicing ribozymes to correct β-globin gene transcript. Gene therapy and gene editing continue to hold promise as a potential curative approach for people with thalassemia. In recent years, genetic-based approaches to thalassemia have begun moving from animal studies into early stage clinical trials involving human subjects. Clinical trials are imminent, it is still not clear how long this novel approach will take to reach the clinic.

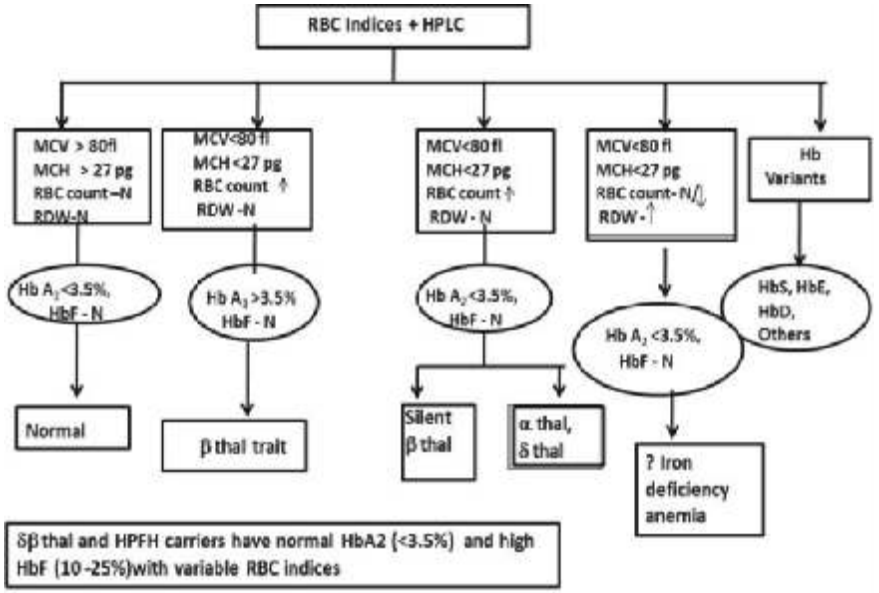
Thalassemia in India

The β thalassemias pose a significant health burden in India. The average prevalence of β thalassemia carriers is 3–4% which translates to 35 to 45 million carriers. Several ethnic groups have a much higher prevalence (4–17%) However, in the absence of National Registries of patients, the exact numbers are not known. Of the 10,000 to 12,000 thalassaemic children born annually in India, very few are optimally managed mainly in urban regions although the Government of India has included the care and

management of patients of thalassemia in their policies. It has been estimated that 2 million units of packed red cells would be needed for transfusion of thalassemia patients in the country. Better management for β thalassemia major patients mainly in urban regions in India with regular and safe blood transfusions and adequate iron chelation allows them to have a better quality of life. However, as they grow older, multi-disciplinary care is required. Although blood is now provided free of cost for patients with hemoglobinopathies and gradually iron chelators are also provided in many states, there are still other expenses for testing, processing and leucodepletion. Thus, majority of the patients do not receive optimum care.

Creating awareness is the key to a successful control program. The Ministry of health along with many NGOs have been conducting education and awareness programs which aim at screening the prospective bride and groom, in fact all youth for thalassemia trait. Identification of carriers is very important, followed by counselling. The National Health Portal of the Ministry of Health and Family Welfare, Govt. of India now provides information on thalassemia for the public and professionals. Yet, there is no formal education on thalassemia in the curriculum for high school children. Intense education on thalassemia from the secondary school level coupled with education of health professionals has been responsible for the success of prevention programmes in the Mediterranean region.

Fig. 1. Screening for carriers of hemoglobinopathies.



Genetic counselling and prenatal diagnosis are important to eliminate the irrational fears among people particularly families at-risk to come to terms with the situation and consequences of the disorder. This will help the couples at-riskto take the right decision for future reproductive choices to avoid the birth of another child with the disease. The crux of genetic counselling is to make the family aware of the genetic disorder, its clinical presentation and severity along with the risk of recurrenceand mortalityassociated with the disease. Communication shouldbe in simple language taking care of the family history, psychologicalissues, ethical issues and cultural and religious challenges.However,there are very few trained counsellors in the country and this aspect needs strengthening.

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