HAESI
HEREDITARY ANGIOEDEMA SOCIETY OF INDIA

3rd National Conference of HAE Society of India (HAESICON)

May 27 - May 28, 2023

Advanced Paediatrics Centre
PGIMER, Chandigarh, India

Supporting Organization

HAEi
HAE International

Awarded 6 Credit hours by Punjab Medical Council (PMC)

Email : haesocietyofindia@gmail.com
Website: www.haesi.in

CSL Behring

Berinert® 500 / 1500

- Treats the fundamental cause of HAE symptoms by replacing the missing protein.[1,2,3]
- Rapid onset of symptom relief regardless of the severity or the location of the HAE attack.[4,5]
- Safe to use in pediatric patients under 12 years.[6,7]

BERINERT®
C1 Esterase Inhibitor, Human
Made in Germany

1. LFrank MM. Complement disorders and hereditary angioedema. J Allergy Clin Immunol. 2010;125:S262-71. **2.** Zuraw BL. Hereditary Angioedema. N Engl J Med. 2008;359:1027-36. **3.** Kaplan AP. The bradykinin-forming cascade and its role in hereditary angioedema. Ann Allergy Asthma Immunol. 2010;104:193-204. **4.** Craig TJ et al. Efficacy of human C1 esterase inhibitor concentrate compared with placebo in acute hereditary angioedema attacks. J Allergy Clin Immunol. 2009;124:801-8. **5.** Craig TJ et al. C1 esterase inhibitor concentrate in 1085 Hereditary Angioedema attacks - final results of the I.M.P.A.C.T. 2 study. Allergy. 2011;66:1604-11. **6.** Busse P et al. Safety of C1-inhibitor concentrate use for hereditary angioedema in pediatric patients. J Allergy Clin Immunol Pract. 2017;5(4):1142-45. **7.** Kreuz W et al. Home therapy with intravenous human C1-inhibitor in children and adolescents with hereditary angioedema. Transfusion. 2012;52:100-7.

Abridged Prescribing Information – Berinert®
Berinert® 500Powder and solvent for solution for injection / infusion. Berinert® 1500 Powder and solution for solution for injection. Qualitative and quantitative composition: Active substance: human C1-esterase inhibitor Berinert® 500 contains 500 IU per injection vial. Berinert® 1500 contains 1500 IU per injection vial. The potency of C1-esterase inhibitor is expressed in International Units (IU), which are related to the current WHO Standard for C1-esterase inhibitor products. Berinert® 500 contains 50 IU/ml C1-esterase inhibitor after reconstitution with 10 ml water for injections and 6.5 mg/ml total protein. Berinert® 1500 contains 500 IU/ml C1-esterase inhibitor after reconstitution with 3 ml water for injections and 65 mg/ml total protein **Therapeutic indications:** Hereditary angioedema type I and II (HAE). Treatment and pre-procedure prevention of acute episodes. **Posology:** ***Adults:*** Treatment of acute angioedema attacks: 20 IU per kilogram body weight (20 IU/kg b.w.) Pre-procedure prevention of angioedema attacks: 1000 IU less than 6 hours prior to a medical, dental, or surgical procedure. ***Paediatric Population:*** Treatment of acute angioedema attacks: 20 IU per kilogram body weight (20 IU/kg b.w.). Pre-procedure prevention of angioedema attacks: 15-30 IU/kg b.w.less than 6 hours prior to a medical, dental, or surgical procedure. **Method of administration:** Berinert® is to be reconstituted. The solution is to be administered by slow i.v. injection. Berinert® 500 can also be administered as infusion (4 ml/minute). **Contraindications:** Hypersensitivity to the active substance or to any of the excipients. **Special warnings and precautions:** In patients with known tendency towards allergies, antihistamines and corticosteroids should be administered prophylactically. Stop Berinert® treatment immediately and initiate appropriate treatment if allergic or anaphylactic reactions occur. Patients with laryngeal oedema require particularly careful monitoring with emergency treatment in stand-by. Berinert® contains up to 486 mg sodium (approximately 21 mmol) per 100 ml solution. To be taken into consideration by patients on a controlled sodium diet. ***Virus safety:*** When medicinal products prepared from human blood or plasma are administered, the possibility of transmitting infective agents cannot be totally excluded. **Interaction with other medicinal products:** No interaction studies have been performed. **Fertility, pregnancy and lactation:** ***Pregnancy:*** Berinert® should be given to a pregnant woman only if clearly needed. ***Breastfeeding:*** It is unknown whether Berinert® is excreted in human milk, but due to its high molecular weight, the transfer of Berinert® into breast milk seems unlikely. A decision must be made whether to discontinue breastfeeding or to discontinue the Berinert® therapy taking into account the benefit of breastfeeding for the child and the benefit of therapy for the woman. ***Fertility:*** No studies on reproduction and developmental toxicity have been performed in animals and no adverse effects on fertility, pre- and postnatal development are expected in humans. **Effects on ability to drive and use machines:** Berinert® has no or negligible influence on the ability to drive and use machines. **Overdose:** No case of overdose has been reported. **Shelf life:** 36 months. **Special precautions for storage:** Do not store above 30°C. Do not freeze. Keep the vial in the outer carton, in order to protect from light. **Date of PI Revision:** June 2021

PLASMAGEN BIOSCIENCES

PLASMAGEN BIOSCIENCES PRIVATE LIMITED
No 160 KCI Chambers, 2nd floor,
5th Main, Chamrajpet, Bangalore 560018.
Contact Number: 080-42154536

ORGANIZING COMMITTEE

Patron

Prof. Surjit Singh

Professor

Head, Department of Paediatrics and Chief, Allergy Immunology Unit,

Advanced Paediatrics Centre, PGIMER Chandigarh, India

Phone No: +91-9872283832

Email: surjitsinghpgi@rediffmail.com

Organizing Chairperson

Prof. Sunil Dogra

Professor

Department of Dermatology, Venereology and Leprology

PGIMER, Chandigarh, India

Phone No: +91-9592065559

Email: sundogra@hotmail.com

Organizing Secretary

Dr. Ankur Jindal

Assistant Professor of Paediatric Clinical Immunology and Rheumatology

Allergy Immunology Unit, Department of Paediatrics,

Advanced Paediatrics Centre, PGIMER Chandigarh, India

Phone No: +91-959206-5559

Email: ankurjindal11@gmail.com

SUB-COMMITTEES

REGISTRATION
Dr. Deepti Suri
Dr. Gayathri
Dr. Rajni Sharma
Mr. Ravinder Garg
Ms. Gurjit Kaur
Mr. Satish Sharma

CATERING
Dr. Amit Rawat
Dr. Rajni Sharma
Dr. Archan Sil
Dr. Reva Tyagi
Dr. Pallavi Nadig

STAGE MANAGEMENT
Dr. Manpreet Dhaliwal
Dr. Rakesh Pilania
Dr. Prabal Barman
Dr. Sanghamitra Machhua
Ms. Sanchi
Dr. Gayathri

SOUVENIR
Dr. M. Sendhil Kumaran
Dr. Sanghamitra Machhua
Dr. Ridhima Aggarwal
Ms. Sanchi
Dr. Reva Tyagi

ACCOMMODATION AND TRANSPORT
Dr. Vignesh P.
Dr. Munish Arora
Dr. Prabal Barman
Dr. Abarna T.
Dr. Rajni Sharma
Dr. Suprit Basu
Ms. Sanchi
Dr. Pallavi Nadig

HAESI
HEREDITARY ANGIOEDEMA SOCIETY OF INDIA

Executive Board of HAESI

Prof. Sunil Dogra
Founder Member & President

Dr. Amit Rawat
Founder Member & President-Elect

Dr. M. Sendhil Kumaran
Founder Member & Vice President

Dr. Ankur Jindal
Founder Member & General Secretary

Dr. Vinay K.
Founder Member and Joint Secretary

Dr. Deepti Suri
Founder Member & Treasurer

Dr. Anuradha Bishnoi
Founder Member & Joint Treasurer

HAESI
HEREDITARY ANGIOEDEMA SOCIETY OF INDIA

WELCOME MESSAGES

Prof. Surjit Singh

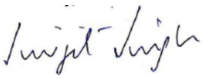
Dear Colleagues,

We are delighted to welcome you to the 3rd National Conference of the Hereditary Angioedema Society of India!

Since its inception in February 2021, the Hereditary Angioedema Society of India has been involved in spreading awareness about HAE in the country. The society has also made attempts in providing better treatment options for patients with HAE in India. As a result, the first line medications for HAE are now available in the country.

I congratulate the Executive Board of the society and the organizing committee for taking up the initiative and organizing this meeting at PGIMER, Chandigarh.

I wish the conference all success.



Prof. Surjit Singh
Head,
Department of Pediatrics and Chief, Allergy Immunology Unit
Postgraduate Institute of Medical Education and Research, Chandigarh

Prof. Sunil Dogra

As president of HAESI, it is my pleasure to welcome you all for HAESICON 2023!

The first conference of HAESI held virtually on 16th May 2021, and 2nd conference held physically on 15th May 2022, which was a grand success on every count. I am immensely pleased that 3rd Conference of HAESI (HAESICON 2023) is being held in physical mode.

I am grateful to all the faculty for making this conference possible. The faculty includes some of the most distinguished names in the field of hereditary angioedema from India and across the globe - Dr. Hilary Longhurst (New Zealand) and Dr. Henriette Farkas (Hungary).

The programme includes didactic lectures and panel discussions on various aspects of hereditary angioedema. In this meeting, we will have Dr. Hilary Longhurst Oration, award paper presentations by residents, fellows and young faculty, and a parallel patient meeting (being organized by Ms. Fiona Wardman, HAE International organization).

While going through details of scientific sessions, it is obvious that they are well conceived and designed. My heartfelt appreciation to team scientific for their untiring efforts. Besides scientific feast, hope you will find some time to enjoy tourist attractions at Chandigarh and nearby hill stations.

Thanks for joining us in the conference – HAESICON 2023!

Warm Regards

Prof. Sunil Dogra
President
HAE Society of India

Dr. Ankur Kumar Jindal

The Hereditary Angioedema Society of India was founded in February 2021 in Chandigarh with an aim to increase awareness and to promote knowledge about HAE amongst physicians, to improve quality of life of patients and bring better treatment options for HAE in India. Over the last 2 year, society has made a substantial contribution towards improving the life of patients with HAE.

We organized the 1st National Conference of HAE Society of India on May 16, 2022 (the international HAE day) and the 2nd National Conference of the HAE Society of India in Chandigarh on May 15, 2022, the country's first-ever physical meeting on HAE, attended by more than 100 delegates nationwide.

We are now hosting the 3rd National Conference of HAE Society of India in Chandigarh on May 27-28, 2023. We are honoured to have Dr. Hilary Longhurst and Dr. Henriette Farkas along with national experts on HAE as faculty for this conference. To encourage young talent, we have invited award paper presentations from fellows, students and young faculty.

Such meetings are often incomplete without involvement of patient groups. We are honored to have Ms. Fiona Wardman and Dr. Shaibal Guha who will lead a parallel patient meeting during the conference.

All this work would not have been possible without the able leadership of Prof. Surjit Singh, Head, Department of Pediatrics and Prof. Sunil Dogra, President, HAE Society of India. I wish to thank Prof. Surjit Singh and Prof. Sunil Dogra for providing continuous support throughout this journey. I also wish to thank other executive board members and organizing team of the 3rd National Conference of HAE Society of India for accomplishing this job very efficiently.

I hope that the delegates would benefit from the proceedings of the conference.

Welcome to Chandigarh - The City Beautiful!

Dr. Ankur Kumar Jindal
Secretary General (HAE Society of India)

HAESI
HEREDITARY ANGIOEDEMA SOCIETY OF INDIA

MEET THE FACULTY

Dr. Aditya Jandial
Senior Research Associate
Department of Clinical Hematology and Medical Oncology,
Postgraduate Institute of Medical Education and Research,
Chandigarh, India

Dr. Aman Gupta
Medical Director and Consultant,
MEDENS Hospital, Panchkula, Haryana

Dr. Amit Rawat
Professor
Allergy Immunology Unit, Department of Paediatrics,
Advanced Paediatrics Centre, PGIMER, Chandigarh, India

Dr. Ankur Jindal
Assistant Professor
Paediatric Clinical Immunology and Rheumatology
Allergy Immunology Unit, Department of Paediatrics,
Advanced Paediatrics Centre, PGIMER, Chandigarh, India

Dr. Anu Maheshwari
Professor
Department of Pediatrics,
Lady Hardinge Medical College and SSK and KSC Hospitals,
New Delhi

Dr. Binay Biswas
Dean
ESIC Medical College Hospital, Bihta, Patna

Dr. Deepti Suri
Professor
Allergy Immunology Unit, Department of Paediatrics,
Advanced Paediatrics Centre, PGIMER, Chandigarh, India

Ms. Fiona Wardman
Co-founder, Director and Chief Executive Officer, HAE Australasia
Director, Treasurer,
Regional Patient Advocate-Asia Pacific and
Chief Regional Patient Advocate, HAE International

Dr. G.P. Thami
Professor and Head
Department of Dermatology
Government Medical College and Hospital, Sector 32
Chandigarh

Dr. Henriette Farkas
Professor
Allergology and Clinical Immunology
Semmelweis University, Budapest, Hungary

Dr. Hilary Longhurst
Senior Medical Officer in Immunology, Auckland
District Health Board Honorary Associate Professorship,
University of Auckland, New Zealand

Dr. Manpreet Dhaliwal
Assistant Professor
Allergy Immunology Unit, Department of Paediatrics,
Advanced Paediatrics Centre, PGIMER, Chandigarh, India

Ms. Pravalika M.
Patient Advocate, HAE India

Dr. Pratap Patra
Additional Professor
Pediatric Clinical Immunology and Rheumatology,
AIIMS, Patna, Bihar

Dr. Rakesh Pilania
Assistant Professor
Department of Paediatrics,
Advanced Paediatrics Centre, PGIMER, Chandigarh, India

Dr. Rashmi Sarkar
Director Professor of Dermatology
Lady Hardinge Medical College and SSK and
KSC Hospitals, New Delhi
President, Indian Academy of Dermatology,
Venereology and Leprology

Dr. Ritambhra Nada
Professor
Department of Histopathology,
PGIMER, Chandigarh, India

Dr. Ruby Pawankar
Executive Director and Immediate Past President, APAAACI
Department of Pediatrics, Nippon Medical School
Tokyo, Japan

Dr. Sagar Bhattad
Consultant
Pediatric Immunologist and Rheumatologist
ASTER CMI Hospitals, Bengaluru, India

Dr. Sameer Gulati
Professor
Department of Medicine, Lady Hardinge Medical College
and SSK and KSC Hospitals, New Delhi

Dr. M. Sendhil Kumaran
Professor
Department of Dermatology, Venereology and Leprology,
PGIMER, Chandigarh, India

Dr. Shaibal Guha
Consultant and Director
Positive Health Centre, Patna Bihar
Founder of HAE India Group

Dr. Srikanta Basu
Director
Professor, Department of Pediatrics, Lady Hardinge Medical College
and SSK and KSC Hospitals, New Delhi

Dr. Sowmya N
Paediatrician
Allergist, and Immunologist, Sanjeevini Specialist Clinic
and Kangaroo Care Hospitals, Bengaluru

Dr. Sukhjot Kaur
Consultant
Department of Dermatology,
Dayanand Medical College & Hospital, Ludhiana

Dr. Sunil Dogra
Professor
Department of Dermatology, Venereology and Leprology,
PGIMER, Chandigarh, India

Dr. Surjit Singh
Professor & Head
Department of Paediatrics,
Chief, Allergy Immunology Unit,
Advanced Paediatrics Centre, PGIMER, Chandigarh, India

Dr. Vignesh Pandiarajan
Associate Professor
Department of Paediatrics,
Advanced Paediatrics Centre, PGIMER, Chandigarh, India

Dr. Vinod Sharma
Assistant Professor
Department of Dermatology, Venereology, and Leprology,
Post Graduate Institute of Medical Education and Research,
Chandigarh

HAESI
HEREDITARY ANGIOEDEMA SOCIETY OF INDIA

SCIENTIFIC PROGRAMME

Scientific Programme

3rd National Conference of Hereditary Angioedema Society of India

Day 1 : May 27, 2023

Time	Title	Speaker	Chairperson
13:00-14:00	**Registration / High Tea**		
14:00-14:10	Introduction and Welcome	Dr. Surjit Singh Dr. Sunil Dogra Dr. Ankur Jindal	
14:15-14:40	Epidemiology of Hereditary Angioedema in the Asia Pacific	Dr. Hilary Longhurst	Dr. Vignesh P. Dr. Binay Biswas
14:45-15:15	Epidemiology of Hereditary Angioedema in India 1. Epidemiology from South India 2. Epidemiology from East India 3. Epidemiology from North - West India	Dr. Sagar Bhattad Dr. Pratap Patra Dr. Ankur Jindal	Dr. Rakesh Pilania Dr. Manpreet Dhaliwal
15:15-15:45	**Tea / Coffee Break**		
15:45-16:10	Clinical mimics of HAE : Case based scenario	Dr. Pallavi Nadig Dr. Keertana Muthukrishnan/ Dr. Ridhima Aggarwal Dr. Lorie Gandhi Dr. Munish Arora	Dr. M. Sendhil Kumaran Dr. Ankur Jindal
16:15-16:40	Ways and means and the importance of setting up a National registry for HAE	Dr. Hilary Longhurst	Dr. Arif Ahmed Dr. Aman Gupta
16:45-17:10	An overview of activities of HAE International (HAEi)	Ms. Fiona Wardman	Dr. Shaibal Guha Ms. Pravalika M.
17:15-17:40	HAE in Asia Pacific Region : Challenges and opportunities	Dr. Ruby Pawankar Dr. Ankur Jindal	Dr. Sagar Bhattad Dr. Anu Maheshwari

Focussed Group Meeting 19:00-20:00; Faculty Dinner : 20:00 onwards

Day 2 : May 28, 2023

Time	Title	Speaker	Chairperson
09:00-10:00	2nd Dr. Hilary Longhurst Oration 'Management of Pediatric patients with hereditary angiodema'	Dr. Henriette Farkas	Dr. Sunil Dogra Dr. Ankur Jindal
10:00-10:25	Oral paper presentation	Dr. Archan Sil Dr. Suprit Basu Dr. Rajni Sharma	Dr. Henriette Farkas Dr. Rashmi Sarkar Dr. Sameer Gulati
10:30-11:00	**Tea / Coffee Break**		
11:00-11:30	**Poster Walk**		
11:30-11:55	Laboratory diagnosis of HAE	Dr. Amit Rawat	Dr. Anuradha Bishnoi Dr. Vinod Sharma
12:00-12:25	Clinicopathologic conference 'A case of laryngeal edema'	Dr. Aditya Jandial Dr. Ritambhra Nada	Dr. Deepti Suri Dr. Srikanta Basu
12:30-12:25	On-demand treatment for HAE: an update? **(Sponsored by CSL Behring)**	Dr. Henriette Farkas	Dr. GP Thami Dr. Sukhjot Kaur
13:00-13:55	Panel discussion How to effectively utilize available resources for diagnosis and management of HAE in India **(Sponsored by Takeda)**	Dr. Hilary Longhurst Dr. Henriette Farkas Dr. Sunil Dogra Dr. Ankur Jindal	
13:55-14:00	Valedictory		

14:00 onwards Lunch

HAESI
HEREDITARY ANGIOEDEMA SOCIETY OF INDIA

ORAL ABSTRACTS

OA 1- Screening for Type II Hereditary Angioedema – the "Poor Man's C1-inhibitor function"

AUTHORS:

Archan Sil[1], Ankur Kumar Jindal[2], Valerie Chiang[3], Prabal Barman[1], Sanchi Chawla[4], Amit Rawat[5], Surjit Singh[6], Elaine YL Au[3], Philip H. Li[7]

AFFILIATIONS:

Senior Resident, Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatrics Centre, Post Graduate Institute of Medical Education and Research, Chandigarh, India- 160012

[2] Assistant Professor, Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatrics Centre, Post Graduate Institute of Medical Education and Research, Chandigarh, India- 160012

[3] Division of Clinical Immunology, Department of Pathology, Queen Mary Hospital, Hong Kong

[4] PhD Scholar, Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatrics Centre, Post Graduate Institute of Medical Education and Research, Chandigarh, India- 160012

[5] Professor, Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatrics Centre, Post Graduate Institute of Medical Education and Research, Chandigarh, India- 160012

[6] Professor and HOD, Department of Pediatrics, Advanced Pediatrics Centre, Post Graduate Institute of Medical Education and Research, Chandigarh, India- 160012

[7] Division of Rheumatology and Clinical Immunology, Department of Medicine, Queen Mary Hospital, University of Hong Kong, Hong Kong

Background- Hereditary angioedema (HAE) is an autosomal dominant disorder characterized by recurrent episodes of angioedema mediated by bradykinin. Most cases of HAE are caused by pathogenic variants in SERPING1 gene. Patients are classified into Type I [C1-inhibitor (C1-INH) deficiency] and Type II [abnormalities in C1-INH function]. Functional assessment of C1-INH is not available in most of the centers. It has been observed that Type II patients seemed to have paradoxically elevated C1-INH levels which may be a useful adjunct to detecting abnormal C1-INH function. This study was therefore performed to assess the diagnostic performance of elevated C1-INH in diagnosing Type II HAE.

Materials and methods- Laboratory and genetic profile of all patients (in Hong Kong and China) with confirmed Type II HAE (as of March 2023) were analyzed. Diagnosis was confirmed by either persistent low C1-INH function and/or pathogenic SERPING1 mutation. All patients had C4 and C1-INH levels quantified by nephelometry. Ethnic-matched individuals without HAE were selected as healthy controls.

Results- There were total 31 patients (14 Chinese and 17 Indian) with Type II HAE and 31 age and sex matched healthy controls. Median age at onset of symptoms and median age at diagnosis were 14.5 (range: 5-25 years) and 40 years (range: 5-77 years) respectively. Median delay in diagnosis was 20.5 years (range: 1-53 years). Male to female ratio was 1:1.6. The sensitivity of low/absent C4 for HAE was 90.3%. Overall, 77.4% (24/31) of Type II HAE patients had elevated C1-INH levels compared to 38.7% (12/31) controls; OR=2.00 (95% CI 1.34-2.98) p=0.017. C1-INH levels among Type II HAE patients were significantly higher than controls (1.97±0.20 vs. 0.94±0.06, p<0.001). A pathogenic variant in exon 8 of SERPING1 gene (c.1396 C>T, p.Arg466Cys) was identified in 30 patients.

Conclusion- Low C4 and presence of elevated C1-INH level may be considered as a screening tool for Type II HAE or the "poor man's C1-INH function".

OA 2-Clinical profile of pediatric hereditary angioedema: our experience from North India

AUTHORS:

Suprit Basu[1], Ankur Kumar Jindal[1], Prabal Barman[1], Reva Tyagi[1], Sanchi Chawla[1], Anit Kaur[1], Sanghamitra Machhua[1], Isheeta Jangra[1], Sendhil M Kumaran[2], Vinay K[2], Anuradha Bishnoi[2], Sunil Dogra[2], Saniya Sharma[1], Manpreet Dhaliwal[1], Deepti Suri[1], Amit Rawat[1], Surjit Singh[1]

AFFILIATIONS:

[1]Pediatric Allergy Immunology Unit, Department of Pediatrics, Postgraduate Institute of Medical Education and Research, Chandigarh, India – 160012
[2]Department of Dermatology,Venerology and Leprology; Postgraduate Institute of Medical Education and Research, Chandigarh, India – 160012

Background: Hereditary angioedema (HAE) is a rare genetic disorder. There are limited data on children with HAE especially from developing countries.
Objective: This study was carried out to analyse the clinical manifestations and laboratory features of children who were diagnosed to have HAE prior to the age of 18 in a tertiary care centre in North India
Methods: Data of children with HAE were retrieved from medical records of patients registered in the Paediatric Immunodeficiency Clinic, Postgraduate Institute of Medical Education and Research, Chandigarh, India.
Results: In this study, we included 59/200 patients (30 male and 29 female) from 42 families diagnosed with HAE before the age of 18 years. The median age at onset of symptoms was 6.1 years (range 1-17 years) and the median age at diagnosis was 10.7 years (range 1.5-18 years) with a median delay in diagnosis of 4.8 years (range 0-16 years). Family history was present in 42/59 (71.2%). Swelling over the face (eyelids and/or lips) was the commonest presentation 83.1% (49/59) followed by extremities and genitalia (Table 1). Laryngeal edema was seen in 44.1% (26/59) cases. Abdominal symptoms were noted in 50.1% of patients. Mean duration of follow-up is 48 months. Type I HAE was seen in 56 patients, and 3 had type-II HAE. Prodromes were seen in 37.3% (22/59) with itching being the most common. Triggers were noted in 55.9% (33/59) patients and trauma was the commonest trigger followed by stress. Six patients reported the death of family members due to HAE. Older age of onset of symptoms, delay in diagnosis and abdominal symptoms were significantly more in adults compared to Pediatric HAE patients. (Table-2). Due to a lack of first-line medications, patients from our cohort received fresh frozen plasma as emergent therapy and tranexamic acid and stanozolol as prophylactic agents. Only 1 patient received C1-esterase inhibitor.
Conclusion: This is one of the largest cohorts of pediatric HAE from a developing country and shows a median delay of 4.8 years for diagnosis of the disease. Hence patient awareness about this rare disease is required.

Table 1: Clinical features of pediatric HAE patients

Clinical features	Number of patients (n=59)
Asymptomatic	3 (5.1%)
Swelling over face (lips and/or eyelids)	49 (83.1%)
Swelling of extremities	45 (76.3%)
Swelling of genitalia	12 (20.3%)
Laryngeal edema	26 (44.1%)
Abdominal pain	30 (50.1%)
Tongue swelling	11 (18.6%)
Family history	42 (71.2%)
Prodromes	22 (37.3%)
Triggers	33 (55.9%)
Type I	56 (94.9%)

Table 2: Clinical features of pediatric HAE patients

Characteristics	Pediatric HAE (n=59)	Adult HAE (n=141)	p value
Male: female	30:29	79:62	0.5
Median age at onset (years)	6.1	17	**<0.05**
Delay in diagnosis (years)	4.8	21	**<0.05**
Facial swelling	49	109	0.36
Extremity swelling	45	104	0.35
Swelling of genitalia	12	33	0.32
Laryngeal edema	26	78	0.07
Abdominal symptoms	30	90	**0.04**
Tongue swelling	11	34	0.19
Prodromes	22	48	0.33
Triggers	33	81	0.42

OA 3-Quality of life in patients with hereditary angioedema correlates with angioedema control: our experience at Chandigarh, India.

AUTHORS:

Rajni Sharma[1], Ankur Kumar Jindal[1], Reva Tyagi[1], Sangeetha Siniah[2], Harshita Umesh[1], Prabal Barman[1], Archan Sil[1], Deepti Suri[1], Vignesh P[1], Amit Rawat[1], Sunil Dogra[3], Surjit Singh[1]

AFFILIATIONS:

[1]Allergy Immunology Unit, Department of Pediatrics, Advanced Pediatrics Centre, PGIMER Chandigarh, India
[2]Hospital Tunku Azizah Women and Children Hospital, Kuala Lumpur
[3]Department of Dermatology and Venereology, PGIMER Chandigarh, India

Background: Lack of first line medications for treatment of HAE in India may lead to poor disease control and add to the poor quality of life.
Objective: To assess the quality of life of patients with HAE using AE QoL scale and identify the factors associated with impaired quality of life.
Methods:
Results: 135 HAE patients (18–80) with a mean age of 40.93 were included in the study (Years). Mean ages at disease onset and diagnosis were 15.7 and 34.9 years, respectively. Most of them had recurring swelling (97.8%), stomach symptoms (65.9%), laryngeal edema (53.3%), and tongue swelling (20.7%). About one-fourth (24.4%) of all individuals reported no QoL impairment, 13.3% mild, 23.7% moderate impairment, and 38.5% significantly damaged QoL. A comparison of disease control indicates that 63.7% of patients reported having their diseases under poor control. 14.3% of them experienced mild, 18.4% moderate, and 67.3% significantly deteriorated quality of life. Disease prevention and QoL were negatively correlated. Patients who had poor illness control rated their quality of life substantially lower than those who had good disease control (p 0.002). Less than one fifth (18.5%) expressed significant anxiety about the unpredictable nature of sickness. One tenth of study participants (9.5%) indicated that their disease was very well controlled with therapy in the previous month, while one fifth of participants (20.7%) reported having no control over their sickness.
Conclusion: This is the first study to report quality of life in patients with HAE in India. Approximately 2/3rd patients have poor quality of life and it negatively correlates with disease control.

HAESI
HEREDITARY ANGIOEDEMA SOCIETY OF INDIA

POSTER ABSTRACTS

PA 1-Use of C1 inhibitor analogue in patients with HAE: our experience from North India

AUTHORS:

Authors: Reva Tyagi[1], Ankur Kumar Jindal[1], Suprit Basu[1], Prabal Barman[1], Sanchi Chawla[1], Anit Kaur[1], Sanghamitra Machhua[1], Isheeta Jangra[1], Sendhil M Kumaran[2], Vinay K[2], Anuradha Bishnoi[2], Sunil Dogra[2], Saniya Sharma[1], Manpreet Dhaliwal[1], Deepti Suri[1], Amit Rawat[1], Surjit Singh[1]

AFFILIATIONS:

[1]Pediatric Allergy Immunology Unit, Department of Pediatrics; Postgraduate Institute of Medical Education and Research, Chandigarh, India – 160012[2]
Department of Dermatology, Venerology and Leprology; Postgraduate Institute of Medical Education and Research, Chandigarh, India – 160012

Background: Hereditary angioedema (HAE) is a rare genetic disorder. This is the first study from India describing the use C1 inhibitor (INH) in patients with HAE.
Objective: This study describes the use of C1INH in patients in a tertiary care centre in North India
Methods: We reviewed the medical records of all patients diagnosed to have HAE in the Pediatric Immunodeficiency Clinic, Department of Pediatrics, Postgraduate Institute of Medical Education and Research, Chandigarh, India. Response and the factors determining response to C1INH analogue was analysed in patients in detail.
Results: Out of 200 patients, we describe use of C1 INH analogue in 6 patients and total use of 13 times. Out of these 6 patients, 2 were male and 4 were female with age ranging from 13 to 30 years. The delay in diagnosis in these patients ranged from 4 years to 23 years. All 6 patients received C1INH analogue for facial swelling, laryngeal edema and abdominal pain. In all these patients there was improvement of symptoms in the form of decrease in the intensity of swelling after receiving C1INH analogue with the promptness of improvement related to the severity and duration of swelling inversely. 2 patients also received Icatibant (bradykinin receptor antagonist) (13 year old male and 39 year old female) as they not show improvement in symptoms within 30 minutes of receiving C1INH analogue.
Conclusion: HAE is associated with significant morbidity and mortality and the use of C1 inhibitor analogues is a new ray of hope in resource limited countries like India.

PA 2- Clinical profile of hereditary angioedema from a community allergy clinic in India

AUTHORS:

M. Ahmed[1]

AFFILIATIONS:

[1]Allermy Center For Allergy And Chest Diseases, Hyderabad, India

Background

Hereditary angioedema (HAE) can be disastrous if u undiagnosed and managed in time. Although a high prevalence is predicted in India, very few cases have been diagnosed. In a retrospective study, we present HAE cases identified in our Community Allergy clinic in last 6 months.

Method

The case records of 8 patients based on clinical presentations and laboratory Reports with due consent from patients are presented. C1 esterase inhibitor (C1 INH) was measured using nephelometry. The relevant data was entered into Microsoft Excel worksheet and analyzed.

Results

A total of 8 patients were diagnosed having HAE with low C1 INH levels in all. The median age at diagnosis was 15 years (range 8 -27). Family history of HAE was seen in 4 cases i.e., 50% and only 1 reported death in the family with undiagnosed HAE like disease. Orofacial oedema was the most common (100%) presentation. Also 3 cases had extremities edema with none of them having abdominal symptoms. All patients were put on tranexamic acid out of which 4 (50%) were taking it irregularly and 2(25%) refused. All were notified about C1 INH inhibitor injection, and all refused the treatment because of cost.

Conclusion

In a short span of six months, we could diagnose 8 HAE cases with consultation regarding the new treatment option available to patients/caregivers. However, spreading awareness about HAE and its diagnosis and management is need of the hour.

PA 3- Delay in diagnosis is the commonest proximate reason for mortality in hereditary angioedema: our experience at Chandigarh, India

AUTHORS:

Prabal Barman[1], Ankur Kumar Jindal[1], Reva Tyagi[1], Suprit Basu[1], Rajni Sharma[1], Sanchi Chawla[1], Sanghamitra Machhua[1], Anit Kaur[1], Rahul Tyagi[1], Isheeta Jangra[1], Sendhil M Kumaran[2], Vinay K[2], Anuradha Bishnoi[2], Sunil Dogra[2], Manpreet Dhaliwal[1], Saniya Sharma[1], Deepti Suri[1], Amit Rawat[1], Surjit Singh[1]

AFFILIATIONS:

[1]Pediatric Allergy Immunology Unit, Department of Pediatrics, Post Graduate Institute of Medical Education and Research, Chandigarh, India-160012
[2]Department of Dermatology, Venereology and Leprology, Post Graduate Institute of Medical Education and Research, Chandigarh, India-160012

Background:
Hereditary angioedema (HAE) is a rare autosomal disorder characterized by non-pruritic subcutaneous/submucosal swellings/oedema (1). Laryngeal oedema is a potentially life-threatening complication (2). Herein, we report our experience with mortality and associated factors of death in patients with HAE.

Methods:
A retrospective review of patients diagnosed to have HAE from Pediatric Immunodeficiency Clinic of Post Graduate Institute of Medical Education and Research, Chandigarh between January 1996 and August 2022 was conducted. Families with HAE who had reported death of at least one family member/ relative because of laryngeal edema were studied in detail.

Results:
Of the 65 families (180 patients), 16 reported the death of at least 1 family member/relative because of laryngeal edema (total 36 deaths). Thirteen families had type-1 HAE and 3 had type-2 HAE. Median age at death (n=31/36) was 38 years (range:17-55 years). At the time of death, at least 1 other family member had angioedema episodes, however, the diagnosis could not be established.

One patient died during follow-up and another patient opted for medical termination of pregnancy when her fetus was found to have the same pathogenic variant.

A comparison of clinical manifestations between families who reported death because of laryngeal edema and families who reported no death showed no significant difference with respect to type of HAE, presence of laryngeal edema, abdominal symptoms and tongue swelling in at least one family member, and molecular defect (Table 1).

Conclusion:
Mortality is still a concern for patients with HAE in India. Laryngeal edema is commonest cause. Delay in diagnosis is the commonest reason for mortality. Awareness and appropriate emergency care need to be prioritized to reduce mortality associated with laryngeal edema with HAE.

PA 4- Interesting case of hereditary angioedema

AUTHORS:

Sowmya Arudi Nagarajan[1] , Harsha N S[2]

AFFILIATIONS:

[1]Department of Allergy and Immunology, Mallige Hospital, Bengaluru, India
[2]Department of Internal Medicine, Bhagawan Mahaveer Jain Hospital, Bengaluru, India

A 37-year-old man presented with 8 years history of recurrent episodes of non-bilious vomiting preceded by abdominal pain which was severe and non-radiating. There was no history of fever, diarrhea, cough or night sweats.Initially he used to get the episodes once in 6 months while now it was occurring almost every month. On examination, his vitals were found to be normal on most of the occasions though twice he had hypotension. Abdomen was found to be soft; sometimes with tenderness and vague abdominal fullness without any organomegaly. These things lead to him being treated as acute gastroenteritis symptomatically; relieving his symptoms over 1-2 days on most of the occasions. Two upper GI endoscopies done in the past were both normal. Blood lead levels, urine porphobilinogen, tissue transglutaminase IgA levels were normal. CT scan of the abdomen done once had shown long segment jejunal thickening; with proximal dilatation in the left side of the abdomen leading to suspicion of intestinal tuberculosis or Crohn's disease or eosinophilic enteritis which then lead to jejunal biopsy being done. It was essentially normal. The patient had stabilized with symptomatic treatment on almost all the occasions though he also had undergone mostly an unnecessary appendicectomy. This continued till the patient once came to us with a different complaint that of swelling of right upper limb which was non-pitting, non-tender and not associated with urticaria.

Discussion

Hereditary angioedema is an autosomal dominant disease caused by a deficiency in functional C1 inhibitor. Its prevalence is estimated to be approximately 1 case per 50,000 persons. The symptoms typically begin in childhood, worsen around puberty, and persist throughout life, with unpredictable severity. Untreated patients have attacks every 7 to 14 days on average, with the frequency ranging from virtually never to every 3 days. Minor trauma and stress are frequent precipitating factors. The arms, legs, hands, feet and abdomen are the most common sites of the swelling. Over half of them have at least one episode of laryngeal oedema which can be fatal. The optimal treatment of hereditary angioedema includes treatment of acute attacks, short-term prophylaxis to prevent an attack, by using c1 inhibitor concentrates or fresh frozen plasma or the newer agents like Ecallantide(selective inhibitor of kallikrein) and Icatibant(bradykinin B2 receptor antagonist). Long-term prophylaxis can be achieved by using alkylated androgens like danazol, stanozolol or anti-fibrinolytic agents like tranexamic acid and EACA., especially in absence of other approved drugs in India. On questioning further he gave a history of having at least 3-4 such episodes of swelling of limbs which used to vanish in 1-2 days. He also gave a significant history that his mother also had recurrent admissions for abdominal pain and vomiting since almost 30 years. She also had puffiness of face and swelling of limbs on and off which was also self-limiting. Their serum C1 esterase inhibitor levels were done and were found to be much below normal in both the patient and his mother leading to a diagnosis of hereditary angioedema. We started the patients, both the mother and son on Danazol 200mg/day. The patient had significant reduction in the frequency and severity of symptoms in the 5 years follow-up with us since then; whereas his mother is totally asymptomatic since then. Our cases highlight that:

PA 5- Hereditary angioedema causing asphyxiation- a case report

AUTHORS:

Ashfaq Masood[1], Mushtaq A Bhat[1]

AFFILIATIONS:

Department of Neonatology & Paediatrics Sher I Kashmir Institute Of Medical Sciences, Jammu & Kashmir

Background: Hereditary angioedema (HAE) is a rare condition characterised by recurrent episodes of swelling of lips and face without urticaria. Severe cases can lead to laryngeal swelling causing asphyxiation.

Objective: We report a 13 year old girl , 2nd in birth order, born of consanguineous marriage, who presented with recurrent episodes of swelling of face and lips without pruritis needing hospitalisations since 5 years of age. There was a family history of similar illness in father, uncle and sister but with less severity. On her first hospital admission patient was evaluated for the cause of her facial swelling. All laboratory reports were normal except C4 level which was decreased [C4=6.4mg/dl]. In 2020 at the age of 10 years patient presented with episode of facial edema with inspiratory stridor. Patient was managed in the intensive care unit with fresh frozen plasma infusions and intravenous tranexamic acid. Patient improved within 48 hours and swelling subsided and patient was discharged. In November 2021 patient presented to the emergency department with facial swelling and severe respiratory compromise. Attendants gave a history of trivial trauma on forehead preceding the onset of symptoms. Patient was hypoxemic with a SpO2 of 65% and absent breath sounds in the chest. Ambu bag ventilation was unsuccessful and endotracheal intubation was difficult. During intubation patient developed cardiopulmonary arrest and needed CPR. Endotracheal intubation was done with a smaller tube using video laryngoscope. Patient was mechanically ventilated for 4 days. Tracheostomy was needed because of extubation failure. After weaning of ventilator patient showed features of hypoxemic brain insult with seizures, increased muscle tone and encephalopathy. MRI brain could not be done due to financial issues. Patient was discharged with advise of physical rehabilitation. Patient had a poor followup and expired in Jan 2023 at home due to complications of asphyxial brain injury.

HAESI
HEREDITARY ANGIOEDEMA SOCIETY OF INDIA

ACADEMIC PARTNERS

CSL Behring

M/s. CSL Behring

Takeda

M/s. Takeda Pharmaceutical company

INTAS

M/s. Intas Biopharmaceuticals

Email : haesocietyofindia@gmail.com
Website: www.haesi.in