HISTORY OF THE INTERNATIONAL GUIDELINES ON HEREDITARY ANGIOEDEMA (HAE) – A RARE DISEASE

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DISCLOSURES

 I have received consultancy/speaker fees and honoraria from Shire, Swedish Orphan Biovitrum, BioCryst, Pharming Octapharma and CSL Behring, as well as research grants from Shire and CSL Behring

CONCEPTION - THE WORKSHOP

Until 1999, only sporadic lectures were presented at different scientific events on HAE.



RESEARCHERS

PHARMA COMPANIES

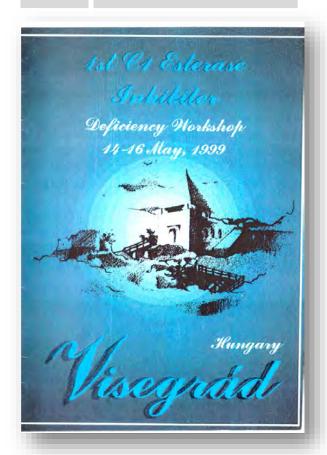


MEDICAL PROFESSIONALS

PATIENTS

MSEGRAD		1999
41	PARTICIPANTS	
12		

41	PARTICIPANTS
13	COUNTRIES
6	SESSIONS
12	SPEAKERS



BUDAPEST 2017 351 PARTICIPANTS 42 COUNTRIES 9 SESSIONS 87 SPEAKERS

VS.



THE MOST IMPORTANT INCREMENTS OF THE WORKSHOP



HAE Organization

News & Events

Resources

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Who is HAEi

HAEi is the international umbrella organization for the world's Hereditary Angioedema (HAE) patient groups.

Our organization is a global non-profit network of patient associations and we are dedicated to raising awareness of C1-inhibitor deficiencies around the world.

We strive to improve time to diagnosis and facilitate access to and reimbursement of life saving HAE therapies, which will enable lifelong health for all patients — no matter where they live.

Prevalence of HAE is still unknown. However, we expect the global prevalence of HAE to be approximately 1 in 30,000, which means that around a quarter million people worldwide suffer from this rare and potentially life threatening deficiency.



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THE METHOD FOR DEVELOPMENT HAE GUIDELINES

A GROUP OF HAE EXPERTS

- drafts a consensus or guideline
- identify, summarize, and evaluate the highest-quality evidence from well-designed and conducted research, as well as the latest data
- presents their recommendations at a *roundtable discussion* held at the Workshop
- the recommendations are voted by the attendees on the spot
- after the Workshop, the manuscript is circulated among the participants
- once all the opinions have been taken into account and harmonized, an expert consensus is published

Rapid publication

Canadian 2003 International Consensus Algorithm for the Diagnosis, Therapy, and Management of Hereditary Angioedema

Tom Bowen, MD, FRCPC, Marco Cicardi, MD (on behalf of PREHAEAT), bx Henriette Farkas, MD PhD, c Konrad Bork, MD, d Wolfhart Kreuz, MD, e Lorenza Zingale, MD, b Lilian Varga, PhD, c Inmaculada Martinez-Saguer, MD, e Emel Aygören-Pürsün, MD, Karen Binkley, MD FRCPC, Bruce Zuraw. MD. Alvin Davis, III, MD, Jacques Hebert, MD, FRCPC, Bruce Ritchie, MD, FRCPC, Jeanne Burnham, Anthony Castaldo, Alejandra Menendez, Istvan Nagy, George Harmat, MD, PhD, ee Christoph Bucher, MD, Gina Lacuesta, MD, FRCPC, P Andrew Issekutz, MD, FRCPC, Richard Warrington, MB, PhD, FRCPC, William Yang, MD, FRCPC, John Dean, MBBS, FRCPC, Amin Kanani, MD, FRCPC, MD, Donald Stark, MD, FRCPC, Christine McCusker, MD, FRCPC, Eric Wagner, PhD, Georges-Etienne Rivard, MD, FRCPC, Eric Leith, MD, FRCPC, Ellie Tsai, MD, FRCPC, Michael MacSween, MD, FRCPC, Dohn Lyanga, MD, FRCPC, Bazir Serushago, MD, FRCPC, aa Art Leznoff, MD, FRCPC, bb Susan Waserman, MD, FRCPC, cc and Jean de Serres, MD^{dd} Calgary, Edmonton, and Claresholm, Alberta, Canada, Milan, Italy, Budapest, Hungary, Mainz and Frankfurt, Germany, Toronto, Ottawa, Oakville, Kingston, Windsor, and Hamilton, Ontario, Canada, La Jolla, Calif, Boston, Mass, Quebec City and Montreal, Quebec, Canada, Annandale, Va, Buenos Aires, Argentina, Zurich, Switzerland, Halifax, Nova Scotia, Canada, Winnipeg, Manitoba, Canada, and Vancouver, British Columbia, Canada

Hereditary angiodema: a current state-of-the-art review, VII: Canadian Hungarian 2007 International Consensus Algorithm for the Diagnosis, Therapy, and Management of Hereditary Angioedema

Tom Bowen, MD, FRCPC: Marco Cicardi, MD; Konrad Bork, MD; Bruce Zuraw, MD; Mike Frank, MD; Bruce Ritchie, MD, FRCPC; Henriette Farkas, MD, PhD, DSc; Lilian Varga, PhD; Lorenza C. Zingale, MD; Karen Binkley, MD, FRCPC; Eric Wagner, PhD; Peggy Adomaitis; Kristylea Brosz, BSc; Jeanne Burnham; Richard Warrington, MB, PhD, FRCPC; Chrystyna Kalicinsky, MD, FRCPC; Sean Mace, MD, FRCPC; Christine McCusker, MD, FRCPC; Robert Schellenberg, MD, FRCPC; Lucia Celeste; Jacques Hebert, MD, FRCPC; Karen Valentine, MD, FRCPC; Man-Chiu Poon, MD, FRCPC; Bazir Serushago, MD, FRCPC; Doris Neurath, BSc, PharmART; William Yang, MD, FRCPC; Gina Lacuesta, MD, FRCPC; Andrew Issekutz, MD, FRCPC; Azza Hamed, MD, FRCPC; Palinder Kamra, MD, FRCPC: John Dean, MBBS, FRCPC: Amin Kanani, MD, FRCPC: Donald Stark, MD, FRCPC; Georges-Etienne Rivard, MD, FRCPC; Eric Leith, MD, FRCPC; Ellie Tsai, MD, FRCPC: Susan Waserman, MD, FRCPC: Paul K, Keith, MD, FRCPC: David Page: Silvia Marchesin: Hilary J. Longhurst, MA, MRCP, PhD, MRCPath: Wolfhart Kreuz, MD, PhD: Eva Rusicke, MD; Inmaculada Martinez-Saguer, MD; Emel Aygören-Pürsün, MD; George Harmat, MD, PhD; George Fust, MD, PhD, DSc; Henry Li, MD, PhD; Laurence Bouillet, MD, PhD; Teresa Caballero, MD, PhD; Dumitru Moldovan, PhD, MD; Peter J. Spath, PhD; Sara Smith-Foltz; Istvan Nagy; Erik W. Nielsen, MD, PhD; Christoph Bucher, MD: Patrik Nordenfelt, MD: and Zhi Yu Xiang, MD

Bowen et al. Allergy, Asthma & Clinical Immunology 2010, **6**:24 http://www.aacijournal.com/content/6/1/24



REVIEW Open Access

2010 International consensus algorithm for the diagnosis, therapy and management of hereditary angioedema

Tom Bowen^{1*}, Marco Cicardi², Henriette Farkas³, Konrad Bork⁴, Hilary J Longhurst⁵, Bruce Zuraw⁶, Emel Aygoeren-Pürsün⁷, Timothy Craig⁸, Karen Binkley⁹, Jacques Hebert¹⁰, Bruce Ritchie¹¹, Laurence Bouillet¹², Stephen Betschel⁹, Della Cogar^{13,14}, John Dean¹⁵, Ramachand Devaraj¹⁶, Azza Hamed¹⁷, Palinder Kamra¹⁷, Paul K Keith¹⁸, Gina Lacuesta¹⁹, Eric Leith²⁰, Harriet Lyons^{13,21}, Sean Mace⁹, Barbara Mako^{13,22}, Doris Neurath²³, Man-Chiu Poon²⁴, Georges-Etienne Rivard²⁵, Robert Schellenberg²⁶, Dereth Rowan^{13,21}, Anne Rowe^{13,27}, Donald Stark²⁶, Smeeksha Sur²⁸, Ellie Tsai²⁹, Richard Warrington³⁰, Susan Waserman¹⁸, Rohan Ameratunga³¹, Jonathan Bernstein³², Janne Björkander³³, Kristylea Brosz^{13,34}, John Brosz^{13,34}, Anette Bygum³⁵, Teresa Caballero³⁶, Mike Frank³⁷, George Fust³, George Harmat³⁸, Amin Kanani²⁶, Wolfhart Kreuz⁷, Marcel Levi³⁹, Henry Li⁴⁰, Inmaculada Martinez-Saguer⁷, Dumitru Moldovan⁴¹, Istvan Nagy⁴², Erik W Nielsen⁴³, Patrik Nordenfelt⁴⁴, Avner Reshef⁴⁵, Eva Rusicke⁷, Sarah Smith-Foltz⁴⁶, Peter Späth⁴⁷, Lilian Varga³, Zhi Yu Xiang⁴⁸

Longhurst et al. Allergy, Asthma & Clinical Immunology 2010, 6:22 http://www.aacijournal.com/content/6/1/22



REVIEW Open Access

HAE international home therapy consensus document

Hilary J Longhurst^{1*}, Henriette Farkas², Timothy Craig³, Emel Aygören-Pürsün⁴, Claire Bethune⁵, Janne Bjorkander⁶, Konrad Bork⁷, Laurence Bouillet⁸, Henrik Boysen⁹, Anette Bygum¹⁰, Teresa Caballero¹¹, Marco Cicardi¹², John Dempster¹³, Mark Gompels¹⁴, Jimmy Gooi¹⁵, Sofia Grigoriadou¹⁶, Ursula Huffer¹⁷, Wolfhart Kreuz¹⁸, Marcel M Levi¹⁹, Janet Long²⁰, Inmaculada Martinez-Saguer²¹, Michel Raguet²², Avner Reshef²³, Tom Bowen²⁴, Bruce Zuraw²⁵





Allergy

REVIEW ARTICLE

Evidence-based recommendations for the therapeutic management of angioedema owing to hereditary C1 inhibitor deficiency: consensus report of an International Working Group

M. Cicardi¹, K. Bork², T. Caballero³, T. Craig⁴, H. H. Li⁵, H. Longhurst⁶, A. Reshef⁷ & B. Zuraw⁸ on behalf of HAWK* (**H**ereditary **A**ngioedema International **W**orking Group)

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Hereditary angioedema with normal C1 inhibitor function: Consensus of an international expert panel

Bruce L. Zuraw, M.D., ^{1,2} Konrad Bork, M.D., ³ Karen E. Binkley, M.D., ⁴ Aleena Banerji, M.D., ⁵ Sandra C. Christiansen, M.D., ^{1,6} Anthony Castaldo, M.P.A., ⁷ Allen Kaplan, M.D., ⁸ Marc Riedl, M.D., ⁹ Charles Kirkpatrick, M.D., ¹⁰ Markus Magerl, M.D., ¹¹ Christian Drouet, Ph.D., ¹² and Marco Cicardi, M.D.

ABSTRACT

A new form of hereditary angioedema (HAE) with normal C1 inhibitor (C1INH) was first described in 2000. The lack of clear diagnostic criteria, the heterogeneity among affected patients, and the varying names given to this disease have led to substantial confusion among both physicians and patients. This study was designed to bring more clarity to the diagnosis and potential treatment of HAE with normal C1INH. An international symposium of experts was convened to review the field and develop consensus opinions that could help clinicians who evaluate and manage these patients. Criteria were developed for the diagnosis of HAE with normal C1INH in patients with recurrent angioedema in the absence of concurrent urticaria. In addition, potential therapeutic strategies are discussed. The consensus criteria developed during this symposium will allow physicians to better diagnose and treat patients with HAE with normal C1INH.

(Allergy Asthma Proc 33:S145-S156, 2012; dot: 10.2500/aap.2012.33.3627)

WAO GUIDELINE

WAO Guideline for the Management of Hereditary Angioedema

Timothy Craig, DO, ¹ Emel Aygören Pürsün, MD, ² Konrad Bork, MD, ³ Tom Bowen, MD, ⁴ Henrik Boysen, ⁵ Henriette Farkas, MD PhD, ⁶ Anete Grumach, MD PhD, ⁷ Constance H. Katelaris, MB BS PhD, ⁸ Richard Lockey, MD, ⁹ Hilary Longhurst, MD, ¹⁰ William Lumry, MD, ¹¹ Markus Magerl, MD, ¹² Immaculada Martinez-Saguer, MD PhD, ² Bruce Ritchie, MD, ¹³ Alexander Nast, MD, ¹² Ruby Pawankar, MD PhD, ¹⁴ Bruce Zuraw, MD, ¹⁵ and Marcus Maurer, MD¹²

Abstract: Hereditary Angioedema (HAE) is a rare disease and for this reason proper diagnosis and appropriate therapy are often unknown or not available for physicians and other health care providers. For this reason we convened a group of specialists that focus upon HAE from around the world to develop not only a consensus on diagnosis and management of HAE, but to also provide evidence based grades, strength of evidence and classification for the consensus. Since both consensus and evidence grading were adhered to the document meets criteria as a guideline. The outcome of the guideline is to improve diagnosis and management of patients with HAE throughout the world and to help initiate uniform care and availability of therapies to all with the diagnosis no matter where the residence of the individual with HAE exists.

Key Words: Hereditary Angioedema, Guidelines, HAE, therapy, management, diagnosis, medications, international

(WAO Journal 2012; 5:182-199)

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Practice paper

International consensus and practical guidelines on the gynecologic and obstetric management of female patients with hereditary angioedema caused by C1 inhibitor deficiency

Teresa Caballero, MD, PhD, a* Henriette Farkas, MD, PhD, DSc, b* Laurence Bouillet, MD, PhD, c* Tom Bowen, MD, d Anne Gompel, MD, PhD, Christina Fagerberg, MD, Janne Bjökander, MD, St, Konrad Bork, MD, h\$ Anette Bygum, MD, h\$ Marco Cicardi, MD, h\$ Caterina de Carolis, MD, h\$ Michael Frank, MD, h\$ Jimmy H. C. Gooi, MD, h\$ Hilary Longhurst, MD, h\$ Immaculada Martínez-Saguer, MD, h\$ Erik Waage Nielsen, MD, h\$ Krystina Obtulowitz, MD, h\$ Roberto Perricone, MD, h\$ and Nieves Prior, MD, h\$ Madrid, Spain, Budapest, Hungary, Grenoble and Paris, France, Calgary, Alberta, Canada, Odense, Denmark, Jönköping, Sweden, Mainz and Frankfurt/Mainz, Germany, Milan and Rome, Italy, Durham, NC, Leeds and London, United Kingdom, Bodø, Norway, and Krakow, Poland





POSITION PAPER

Classification, diagnosis, and approach to treatment for angioedema: consensus report from the Hereditary Angioedema International Working Group

M. Cicardi¹, W. Aberer², A. Banerji³, M. Bas⁴, J. A. Bernstein⁵, K. Bork⁶, T. Caballero⁷, H. Farkas⁸, A. Grumach⁹, A. P. Kaplan¹⁰, M. A. Riedl¹¹, M. Triggiani¹², A. Zanichelli¹ & B. Zuraw¹¹ on behalf of HAWK, under the patronage of EAACI (European Academy of Allergy and Clinical Immunology)*

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ORIGINAL ARTICLE

IMMUNODEFICIENCIES

International consensus on the diagnosis and management of pediatric patients with hereditary angioedema with C1 inhibitor deficiency

H. Farkas¹, I. Martinez-Saguer², K. Bork³, T. Bowen⁴, T. Craig⁵, M. Frank⁶, A. E. Germenis⁷, A. S. Grumach⁸, A. Luczay³, L. Varga¹ & A. Zanichelli¹⁰ on behalf of HAWK¹

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Position Paper

The international WAO/EAACI guideline for the management of hereditary angioedema – the 2017 revision and update

Marcus Maurer ☑, Markus Magerl, Ignacio Ansotegui, Emel Aygören Pürsün,
Stephen Betschel, Konrad Bork, Tom Bowen, Henrik Balle Boysen, Henriette Farkas,
Anete Grumach, Michihiro Hide, Constance Katelaris, Richard Lockey, Hilary Longhurst,
William Lumry, Inmaculada Martinez-Saguer, Dumitru Moldovan, Alexander Nast,
Ruby Pawankar, Paul Potter, Marc Riedl, Bruce Ritchie, Lanny Rosenwasser,
Mario Sánchez-Borges, Yuxiang Zhi, Bruce Zuraw, Timothy Craig

CONSENSUS

Practice paper

International consensus and practical guidelines on the gynecologic and obstetric management of female patients with hereditary angioedema caused by C1 inhibitor deficiency

Teresa Caballero, MD, PhD, ** Henriette Farkas, MD, PhD, DSc, ** Laurence Bouillet, MD, PhD, c** Tom Bowen, MD, d Anne Gompel, MD, PhD, e Christina Fagerberg, MD, Janne Bjökander, MD, Janne Bjökande

CONSENSUS

NEW

International consensus statement on the application of genetics in the management of primary angioedema

ROUND TABLE DISCUSSIONS

11th C1-INHIBITOR DEFICIENCY AND ANGIOEDEMA WORKSHOP 23-26 MAY, 2019, BUDAPEST

