Allergology International 70 (2021) 235–243

Contents lists available at ScienceDirect

Allergology International

journal homepage: http://www.elsevier.com/locate/alit

Original Article

The diagnosis and treatment of hereditary angioedema patients in Japan: A patient reported outcome survey

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ARTICLE INFO

Article history:
Received 7 May 2020
Received in revised form
7 September 2020
Accepted 23 September 2020
Available online 6 November 2020

Keywords:
Consultation behavior
Hereditary angioedema
Japan
Patient survey
QoL

Abbreviations:

C1-INH	C1 inhibitor
HAE	Hereditary angioedema
pdC1-INH	plasma-derived C1 inhibitor concentrate

ABSTRACT

Background: The rate at which patients are accurately diagnosed with hereditary angioedema (HAE), as well as diagnosed patients access to modern treatments differs greatly among countries. Moreover, the severity and burden of HAE on patients have been reported mostly on the basis of physician-reported surveys. To gain insight into the real-world conditions of patients with HAE through a patient-reported survey in Japan and identify any unmet needs.

Methods: A questionnaire was distributed to 121 patients with HAE via a Japanese HAE patient organization during 2016–2017. Responses were collected from 70 patients (57.9%) and subjected to analysis.

Results: The average periods from the initial appearance of symptoms (e.g. edema) to a HAE diagnosis was 15.6 years (min–max, 0–53). Patients visited an average of 4.6 different departments until receiving a definitive diagnosis. The average age at the first visit was 25.6 years (3–73) and at diagnosis 32.8 years (0–73). Patients reported an average of 15.7 (0–100) attacks per year, but only 53.1% of attacks were treated. The days of hospitalization due to severe attacks was 14.3 (0–200) before diagnosis, but these declined to 4.3 (0–50) after diagnosis. In the treatment for attacks, 82% of the patients were treated with the plasma-derived C1 inhibitor concentrate, and 69% of the patients reported experiencing a therapeutic effect.

Conclusions: There is a long gap between first attack and diagnosis of HAE, and the number of non-treated attacks is high in Japan. Steps are needed to improve the diagnostic and treatment environments to address these issues.

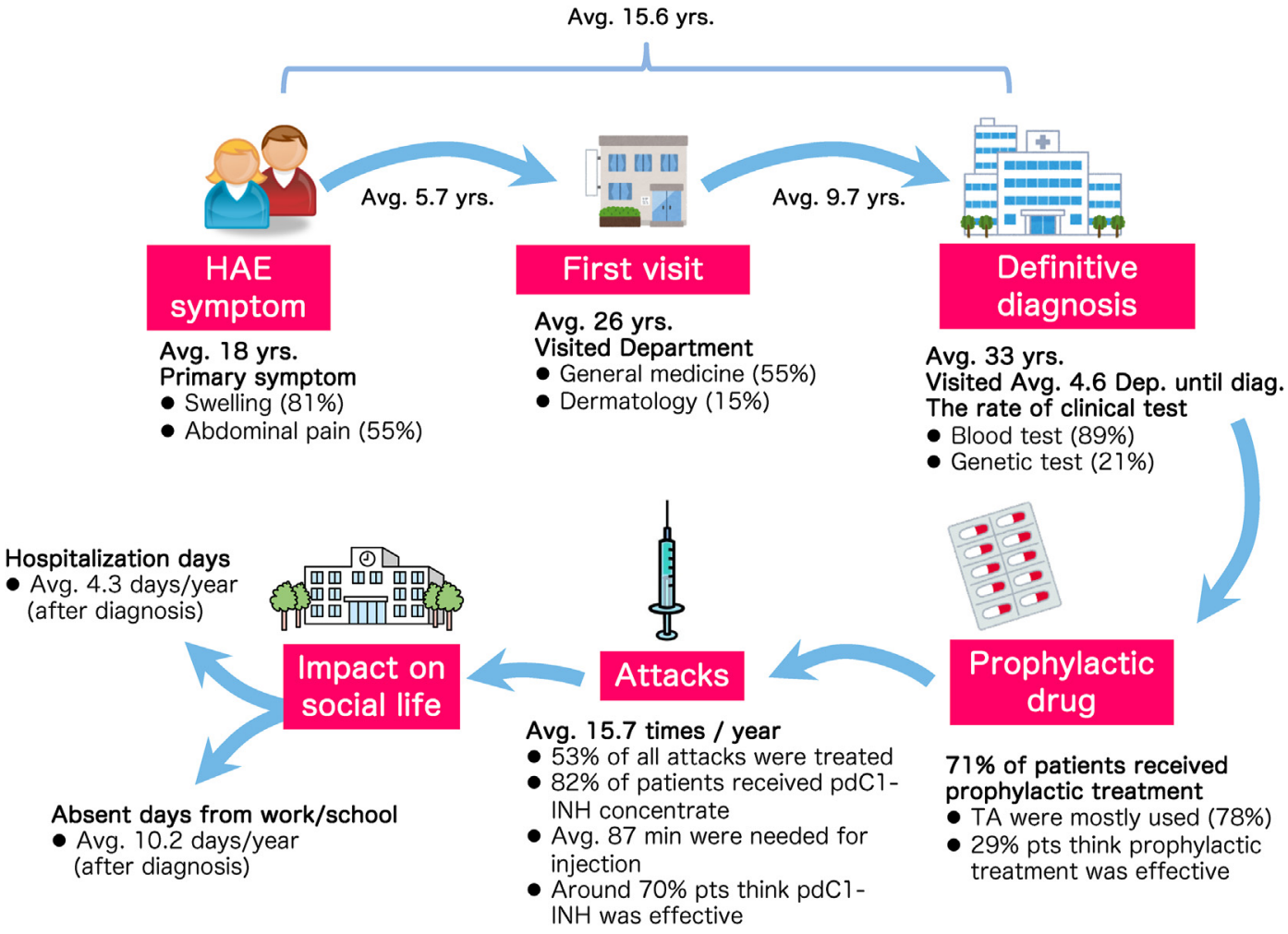

Fig. 8. Graphical summary.

遺伝性血管性浮腫（HAE：エイチ・エー・イー）は、手足の浮腫（むくみ）や腹痛を繰り返す病気です。突然はげしい症状が出ることもあり、また病名のとおり遺伝する病気です。

上記の症状が当てはまる場合には、専門の医療機関の受診が必要です。当院までご相談ください。