

Innovation to collect data for a national registry of patients with hereditary angioedema: the first phase of establishing the dynamic HARPE registry

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BACKGROUND

HARPE, a dynamic registry for hereditary angioedema (HAE):

- Compliant with the **European patient data protection regulations**,
- A source of data for the **European CARE registry**,
- A source of data for **research projects**, after validation by the scientific advisory board.

In rare diseases, the **collection of real-life data is essential** for:

- Understanding the disease** (genetic variants, association with comorbidities),
- Evaluating patient quality of life** (identification of needs),
- Research and development** of new therapies (phase 4 studies, etc.).

Obstacles for traditional data collection:

- Time-consuming** and limited resources, particularly in smaller centers,
- Risk of errors** during data entry,
- Delays** in obtaining an up-to-date, complete database.

Today, we can harness innovation to collect and structure data for rare disease, such as HAE.

METHODOLOGY

The outstanding feature of this project is the use of an **innovative approach** to identify patients, as well as collect and structure data to establish the registry. The data were extracted from the **patients' electronic records** and automatically structured using the Lifen DataLab solution. This solution uses **artificial intelligence natural language processing (AI-NLP)** to structure data.

VARIABLES

A set of **160 variables** were identified, in six domains:

- Patient and disease history (including comorbidities)
- Diagnosis
- Intercurrent events
- General information
- Healthcare management (including details of consultations)
- Treatments

Figure: Lifen DataLab — data collection and structuring tool

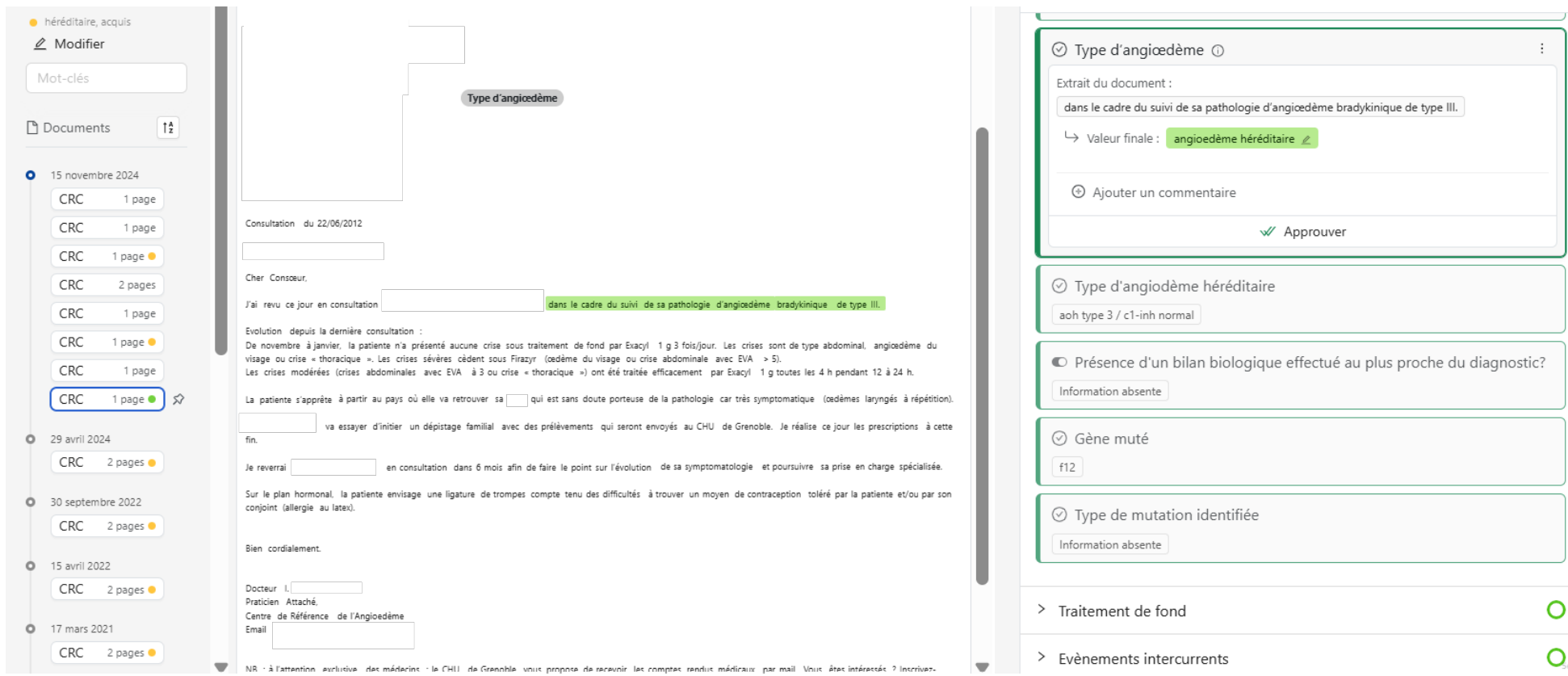


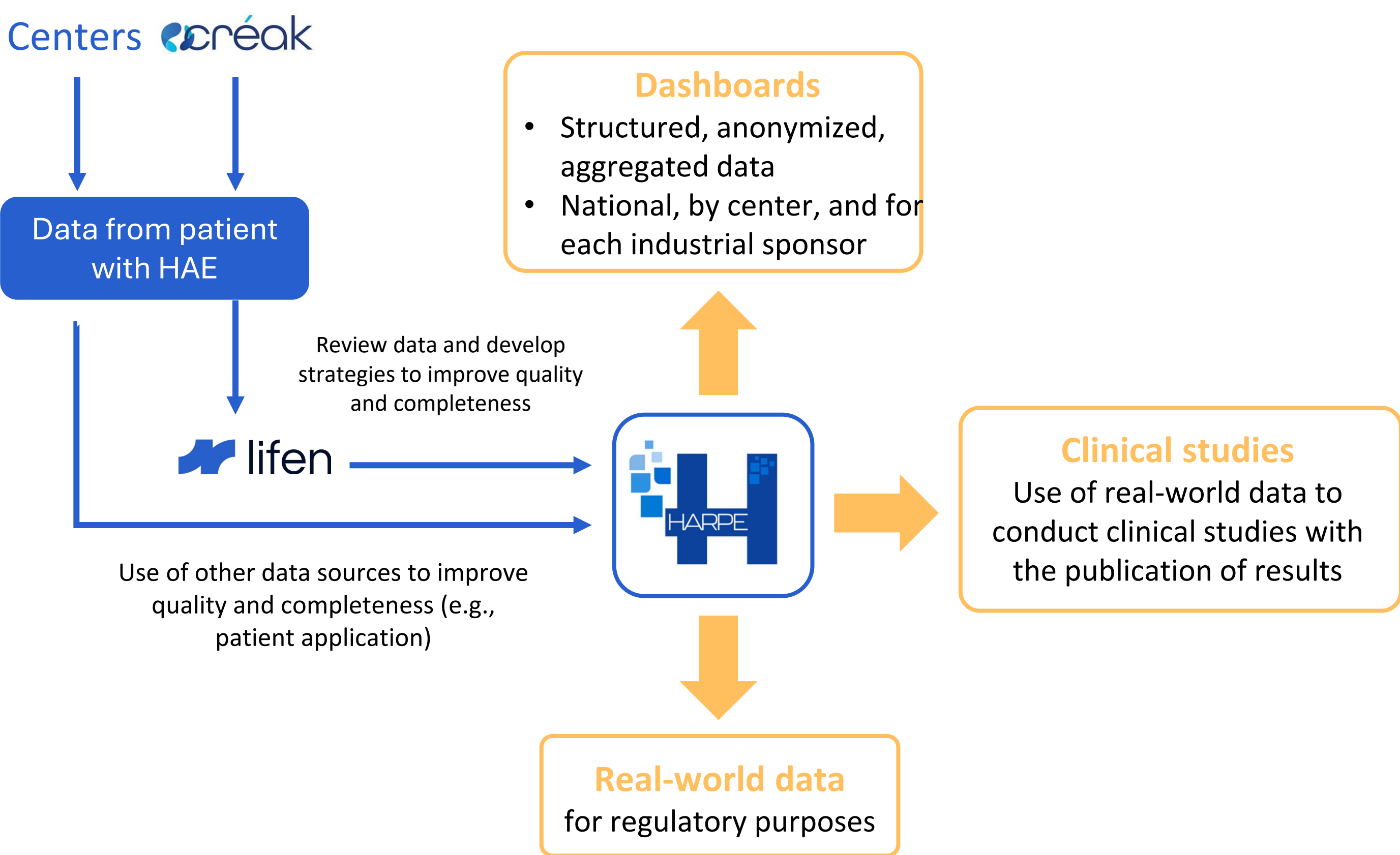
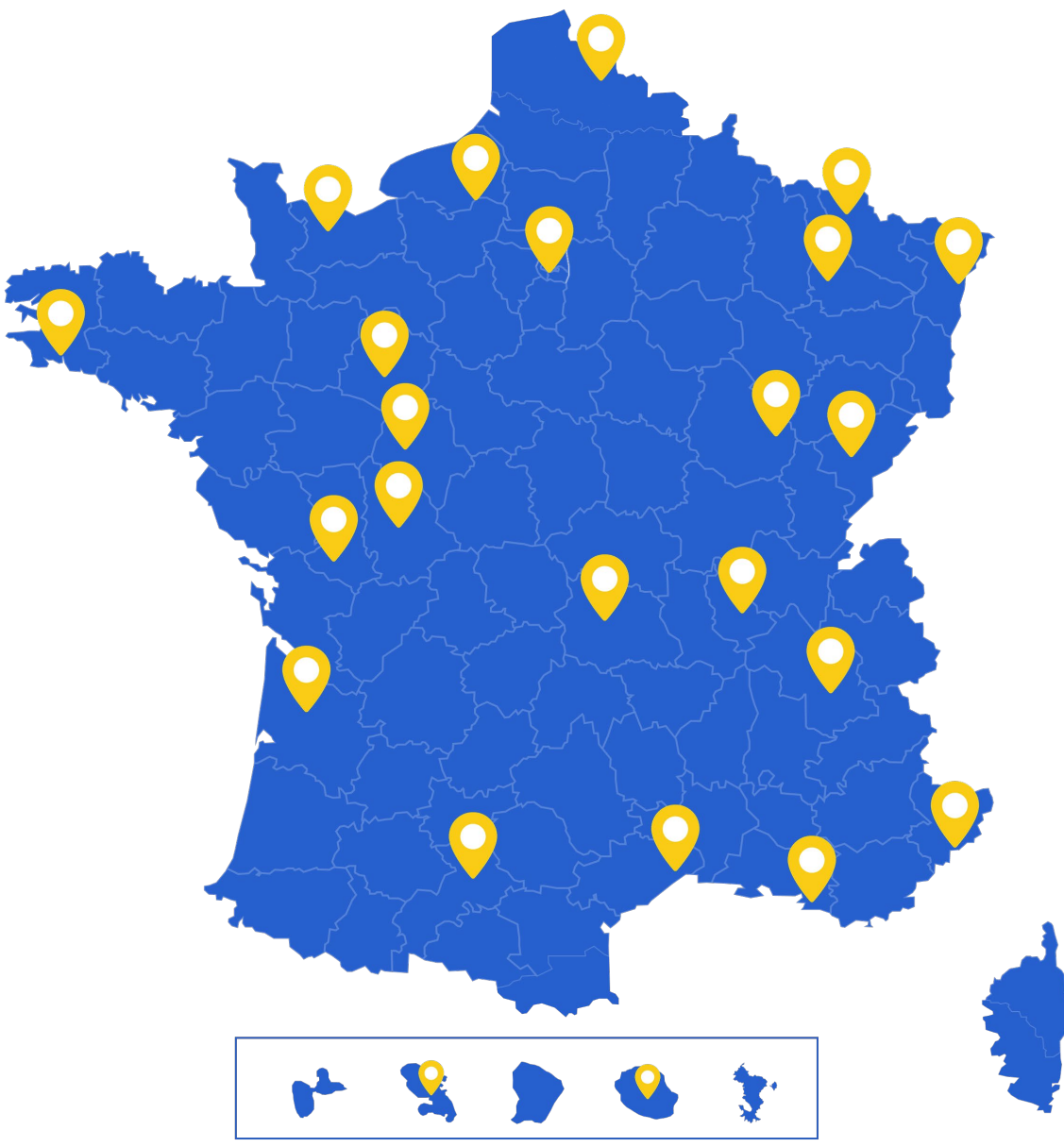
Figure: Design and objective of the HARPE registry

OBJECTIVE

To develop a dynamic registry using AI to collect and structure data.



- National reference center for angioedema:
- Network of 24 centers,
 - Coordinator: Grenoble Alpes University Hospital.
- Among its objectives:
- Provide expert diagnostic and therapeutic care,
 - Advance research



RESULTS

Among patients followed at the Grenoble Alpes University Hospital:

- 198 patients with HAE** were identified by AI analysis of documents in the patient files
- The average completeness rate for the 160 variables was **66.82%**.

Table: Degree of “completeness” and strategies for improving data completeness and quality

Variables	n	Values	Completeness (%)
Age at first HAE attack	198	20.1	19
Median age at confirmed diagnosis	198	26.2	32
Sex : Female	198	65	100
Median duration of follow-up from diagnosis	198	11.1	33
Type of HAE: Type 1	198	129	100
Type of HAE: Type 2	198	8	100
Type of HAE: Type 3	198	53	100
Type of HAE: Type unknown	198	8	100
On-demand treatment ^a	198	34	90
Long-term prophylaxis	198	97	51
Number of patients consulted within 12 months	99	99	100
Number of patients with 1 consultation within 12 months	99	18	100
Number of patients with 2 consultations within 12 months	99	38	100
Number of attacks over 12 months	99	239	100
Number of hospitalizations	35	1	100

^aIn theory, all patients receive on-demand treatment.

NEXT STEPS

The next steps will aim to:

- Confirm data quality** (manual versus AI collection study),
- Extend the data collection** to other centers in the CREAK network,
- Improve the completeness rate**, notably through the addition of a “patient” application to be implemented shortly,
- Provide additional data** on treatments, HAE attacks, and patients' quality of life via the application.

CONCLUSION

This innovative and dynamic registry provides up-to-date real-world data to assess and improve therapeutic outcomes, patient quality of life, standardize practices, and develop scientific and medical research. Once established the registry can be rapidly updated with new patients and additional data. Natural language processing (NLP) is an AI tool capable of achieving these objectives in this rare disease setting.

¹Bork K, Anderson JT, Caballero T, et al. Assessment and management of disease burden and quality of life in patients with hereditary angioedema: a consensus report. Allergy Asthma Clin Immunol. 2021;17(1).

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