

## Incidence and risk factors for clinical neurodegenerative Langerhans cell histiocytosis: a longitudinal cohort study

Submitted by Beatrice Guillaumat on Wed, 01/09/2019 - 14:13

Titre Incidence and risk factors for clinical neurodegenerative Langerhans cell histiocytosis: a longitudinal cohort study

Type de publication Article de revue

Auteur H ritier, S bastien [1], Barkaoui, Mohamed-Aziz [2], Miron, Jean [3], Thomas, Caroline [4], Moshous, Despina [5], Lambilliotte, Anne [6], Mazingue, Fran oise [7], Kebaili, Kamila [8], Jeziorski, Eric [9], Plat, Genevi ve [10], Aladjidi, Nathalie [11], Pacquement, H l ne [12], Galambrun, Claire [13], Brugi res, Laurence [14], Leverger, Guy [15], Mansuy, Ludovic [16], Paillard, Catherine [17], Deville, Anne [18], Pagnier, Anne [19], Lutun, Anne [20], Gillibert-Yvert, Marion [21], Stephan, Jean-Louis [22], Cohen-Aubart, Fleur [23], Haroche, Julien [24], Pellier, Isabelle [25], Millot, Fr d ric [26], Gandemer, Virginie [27], Martin-Duverneuil, Nadine [28], Taly, Val rie [29], H lias-Rodzewicz, Zofia [30], Emile, Jean-Fran ois [31], Hoang-Xuan, Khe [32], Idbaih, Ahmed [33], Donadieu, Jean [34]

Editeur Wiley

Type Article scientifique dans une revue   comit  de lecture

Ann e 2018

Langue Anglais

Date Novembre 2018

Num ro 4

Pagination 608-617

Volume 183

Titre de la revue British journal of haematology

ISSN 1365-2141

Mots-cl s BRAF [35], histiocytosis [36], Langerhans cell histiocytosis [37], neurodegeneration [38]

Résumé en anglais

Neurodegenerative (ND) complications in Langerhans cell histiocytosis (LCH) are a late-onset but dramatic sequelae for which incidence and risk factors are not well defined. Based on a national prospective registry of paediatric LCH patients, we determined the incidence rate of clinical ND LCH (cND-LCH) and analysed risk factors, taking into account disease extent and molecular characteristics. Among 1897 LCH patients, 36 (1.9%) were diagnosed with a cND-LCH. The 10-year cumulative incidence of cND-LCH was 4.1%. cND-LCH typically affected patients previously treated for a multisystem, risk organ-negative LCH, represented in 69.4% of cND-LCH cases. Pituitary gland, skin and base skull/orbit bone lesions were more frequent ( $P < 0.001$ ) in cND-LCH patients compared to those without cND-LCH (respectively 86.1% vs. 12.2%, 75.0% vs. 34.2%, and 63.9% vs. 28.4%). The 'cND susceptible patients' ( $n = 671$ ) i.e., children who had experienced LCH disease with pituitary or skull base or orbit bone involvement, had a 10-year cND risk of 7.8% vs. 0% for patients who did not meet these criteria. Finally, BRAF status added important information among these cND susceptible patients, with the 10-year cND risk of 33.1% if a BRAF mutation was present compared to 2.9% if it was absent ( $P = 0.002$ ).

URL de la notice	<a href="http://okina.univ-angers.fr/publications/ua18568">http://okina.univ-angers.fr/publications/ua18568</a> [39]
DOI	10.1111/bjh.15577 [40]
Lien vers le document	<a href="https://onlinelibrary.wiley.com/doi/abs/10.1111/bjh.15577">https://onlinelibrary.wiley.com/doi/abs/10.1111/bjh.15577</a> [41]
Titre abrégé	Br. J. Haematol.
Identifiant (ID) PubMed	30421536 [42]

---

## Liens

- [1] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32521>
- [2] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32522>
- [3] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32523>
- [4] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32524>
- [5] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32525>
- [6] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32526>
- [7] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=31915>
- [8] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32527>
- [9] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32528>
- [10] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32529>
- [11] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=31905>
- [12] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32530>
- [13] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=31913>
- [14] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32531>
- [15] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32532>
- [16] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32533>
- [17] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32534>
- [18] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32535>
- [19] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32536>
- [20] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32537>
- [21] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32538>
- [22] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32539>
- [23] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32540>
- [24] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32541>

- [25] <http://okina.univ-angers.fr/i.pellier/publications>
- [26] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32542>
- [27] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=30452>
- [28] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32543>
- [29] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32544>
- [30] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32545>
- [31] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=8044>
- [32] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=32546>
- [33] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=30608>
- [34] <http://okina.univ-angers.fr/publications?f%5Bauthor%5D=31925>
- [35] <http://okina.univ-angers.fr/publications?f%5Bkeyword%5D=17326>
- [36] <http://okina.univ-angers.fr/publications?f%5Bkeyword%5D=26944>
- [37] <http://okina.univ-angers.fr/publications?f%5Bkeyword%5D=26943>
- [38] <http://okina.univ-angers.fr/publications?f%5Bkeyword%5D=15466>
- [39] <http://okina.univ-angers.fr/publications/ua18568>
- [40] <http://dx.doi.org/10.1111/bjh.15577>
- [41] <https://onlinelibrary.wiley.com/doi/abs/10.1111/bjh.15577>
- [42] <http://www.ncbi.nlm.nih.gov/pubmed/30421536?dopt=Abstract>

Publié sur *Okina* (<http://okina.univ-angers.fr>)