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Diagnosis and treatment of vulvo-perineal endometriosis: a systematic review

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Charlotte Maillard[1, 2*], Zineb Cherif Alami[3], Jean Squifflet[1, 2], Mathieu Luyckx[2, 4], Pascale Jadoul[1, 2], Viju Thomas[5, 6] and Christine Wyns[2, 7]

[1]Catholic University of Louvain, Belgium
[2]Cliniques Universitaires Saint-Luc, Belgium
[3]Saint Jean Clinic, Belgium
[4]de Duve Institute, Université Catholique de Louvain, Belgium
[5]Department of Medicine, Faculty of Medicine and Health Sciences, Stellenbosch University, South Africa
[6]Tygerberg Hospital, South Africa
[7]Institute of Experimental and Clinical Research, Catholic University of Louvain, Belgium

Objective: To describe the available knowledge on vulvo-perineal endometriosis including its diagnosis, clinical management and recurrence rate.
Methods: We followed the PRISMA guidelines for Systematic Reviews and our study was prospectively registered with PROSPERO (CRD42020202441). The terms "Endometriosis" and "Perineum" or "Vulva" were used as keywords. Cochrane Library, Medline/Pubmed, Embase and Clinicaltrials.gov were searched. Papers in English, Spanish, Portuguese, French or Italian from inception to July 30, 2020 were considered. Reference lists of included articles and other literature source such as Google Scholar were also manually scrutinized in order to identify other relevant studies. Two independent reviewers screened potentially eligible studies according to inclusion criteria.
Results: Out of 539 reports, 90 studies were eligible including a total of 283 patients. Their mean age was 32,7±7,6 years. Two hundred sixty-three (95,3%) presenting with vulvo-perineal endometriosis have undergone either episiotomy, perineal trauma or vaginal injury or surgery. Only 13 patients (4,7%) developed vulvo-vaginal endometriosis spontaneously i.e. without any apparent condition favoring it. The reasons that motivated the patients to take medical advice were vulvo-perineal cyclical pain increasing during menstruations (98,2% of the patients, n=278). Out of the 281 patients for whom a clinical examination was described, 274 patients (97,5%) showed a vulvo-perineal nodule, mass or swelling while 6 presented with bluish cutaneous lesions (2,1%) and 1 with bilateral polyps of the labia minora (0,4%). All but one patients underwent surgical excision of their lesions but only 88 patients (28,1%) received additional hormonal therapy. The recurrence rate was 10,2% (29 patients) considering a median follow-up period of 10 months (based on 61 studies).
Conclusion: Vulvo-perineal endometriosis is poorly reported with only approximately 300 cases identified in the literature since 1923. The limited quality of available studies does not allow to support an evidence-based approach to the diagnosis and treatment of affected patients. However, development of a transnational registry of patients with extra-pelvic endometriosis, as already suggested by Andres et al. (2019), could prove useful to further increase knowledge but also awareness among healthcare professionals and optimize patients' care.

Keywords: Endometriosis, Perineum, Vulva, Episiotomy, Perineal pain, Cyclical pain, Perineal nodule, Extrapelvic endometriosis

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* **Correspondence:** Dr. Charlotte Maillard, Catholic University of Louvain, Louvain-la-Neuve, Belgium, Maillardcv@gmail.com