

ANALYSIS OF THE PREVALENCE OF ORAL CANCER IN PATIENTS WITH DISKERATOSIS CONGENITA: A SYSTEMATIC REVIEW

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Goal: is to present the prevalence of oral cancer in patients with dyskeratosis congenita (CD). Theme: Dyskeratosis congenita (CD) is the first genetic syndrome described among telomeropathies. Dyskeratosis congenita (CD) is commonly diagnosed clinically based on a classic phenotype: 1) oral leukoplakia, 2) nail dystrophy and 3) abnormal skin pigmentation (cutaneous lace reticulate pigmentation). It is commonly associated with bone marrow failure, increased predisposition to malignancies, and a variety of additional somatic features. Dyskeratosis congenita presents marked genetic heterogeneity, as at least 14 genes are responsible for the shortening of telomeres characteristic of this disease. Materials and methods: This is a Systematic Review, in which the research question is structured in the format of the anachronistic PECO, where P = Population of interest (patient or problem to be addressed), E = Exposure (to be considered, for example: risk factor, prognostic factor); C = Comparator (control, for example: non-exposure); O = Outcome (outcome of interest or situation of interest). Therefore, the key question chosen was: What is the prevalence of oral cancer in patients with dyskeratosis congenita? Using the Capes Periódicos, SciELO and Bireme databases, approximately 21 articles were initially selected that address the topic relating the appearance of oral cancer in patients with dyskeratosis congenita (CD). From this quantity, 10 articles were selected for registration; Of these 10 registered articles, 5 were selected to be used as a basis (sentinel), as well as to observe among the characteristics whether they are prospective/retrospective or case reports, and outline eligibility criteria (inclusion or exclusion). Expected results: According to a study presented by John M. Le, Michael T. Kae and Anthony B. Morlandt (2020), almost 70% of patients with CD have oral leukoplakia, a lesion with up to a 34% chance of malignant transformation. Although oral squamous cell carcinoma (OSCC) is the most common malignant transformation of long-standing leukoplakia, a limited number of cases have been reported in patients with CD. Most involved the tongue and oral mucosa.

Key words: Oral Cancer; Dyskeratosis congenita (DC); Prevalence; Oral.

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