Research, Society and Development, v. 14, n. 3, e3614348388, 2025
(CC BY 4.0) | ISSN 2525-3409 | DOI: http://dx.doi.org/10.33448/rsd-v14i3.48388

How scientific evidence can contribute to improve the diagnosis and comprehensive care of patients with TEMPI syndrome: A scoping review protocol

Como as evidências científicas podem contribuir para melhorar o diagnóstico e o cuidado integral de pacientes com síndrome de TEMPI: Um protocolo de revisão de escopo

Cómo la evidencia científica puede contribuir a mejorar el diagnóstico y la atención integral de los pacientes con síndrome de TEMPI: Un protocolo de revisión de alcance

Received: 02/24/2025 | Revised: 03/07/2025 | Accepted: 03/08/2025 | Published: 03/13/2025

Vivian Menezes Irineu
ORCID: https://orcid.org/0000-0003-4024-0271
Universidade Municipal de São Caetano do Sul, Brazil
E-mail: vivian.irineu@online.uscs.edu.br
Maria Júlia Pinheiro Fernandes
ORCID: https://orcid.org/0009-0006-0769-422X
Universidade Municipal de São Caetano do Sul, Brazil
E-mail: maria.fernandes2@uscsonline.com.br

Abstract
Objective: To document the planning and methodological processes of a scoping review protocol aimed at mapping the current scientific knowledge on TEMPI Syndrome. Introduction: TEMPI Syndrome is a rare, acquired, and multisystemic disease characterized by telangiectasias, elevated erythropoietin levels and erythrocytosis, monoclonal gammopathy, perinephric fluid collections, and intrapulmonary shunting. Justification: Due to its rarity, we hypothesize that the lack of awareness regarding this syndrome may lead to misdiagnosis or delayed diagnosis. A systematic literature review can contribute to increasing knowledge and dissemination of information on the topic. Methodology: The protocol was registered in the Open Science Framework and structured following the five-stage methodological framework by Arksey & O'Malley (2005), in accordance with the Joanna Briggs Institute guidelines. The review is guided by the question: "How can the available scientific evidence contribute to improving the diagnosis and comprehensive care of patients with TEMPI Syndrome?" A systematic search will be conducted in the Medical Literature Analysis and Retrieval System (MEDLINE) and in the Biblioteca Virtual em Saúde (BVS), without time restrictions. Gray literature will be retrieved using Google Scholar. Two independent reviewers will screen studies for eligibility. Results: Findings will be reported in accordance with the PRISMA-ScR (Preferred Reporting Items for Systematic Reviews and Meta-Analyses extension for Scoping Reviews) and presented in tabular and narrative formats aligned with the study objectives and research question.
Keywords: Plasma cell dyscrasia; Erythrocytosis; Monoclonal gammopathies; Research protocol; TEMPI Syndrome.

Resumo
Objetivo: Documentar o planejamento e os processos metodológicos de um protocolo de scoping review visando mapear o conhecimento científico atual sobre a Síndrome TEMPI. Introdução: A Síndrome TEMPI é uma doença rara, adquirida e multissistêmica caracterizada por telangiectasias, níveis elevados de eritropoietina e eritrocitose, gamopatia monoclonal, coleções de líquido perinéfrico e shunt intrapulmonar. Justificativa: Devido à sua raridade, levantamos a hipótese de que a falta de conhecimento sobre esta síndrome pode levar a erros de diagnóstico ou atraso no diagnóstico. Uma revisão sistemática da literatura pode contribuir para aumentar o conhecimento e a divulgação de informações sobre o tema. Metodologia: O protocolo foi registrado no Open Science Framework e estruturado seguindo o referencial metodológico de cinco etapas de Arksey & O'Malley (2005), de acordo com as diretrizes do Joanna Briggs Institute. A revisão é norteada pela questão: "Como as evidências científicas disponíveis podem contribuir para melhorar o diagnóstico e o cuidado integral dos pacientes com Síndrome TEMPI?" Será realizada uma busca sistemática no Medical Literature Analysis and Retrieval System (MEDLINE) e na Biblioteca Virtual em Saúde (BVS), sem restrição de tempo. A literatura cinzenta será recuperada usando o Google Scholar. Dois revisores independentes avaliarão os estudos quanto à elegibilidade. Resultados: Os resultados serão relatados de acordo com o PRISMA-ScR (Preferred Reporting Items for Systematic Reviews and Meta-Analyses extension for Scoping Reviews) e apresentados em formatos tabulares e narrativos alinhados com os objetivos do estudo e a questão da pesquisa.
Palavras-chave: Discrasia de células plasmáticas; Eritrocitose; Gamopatias monoclonais; Protocolo de pesquisa; Síndrome de TEMPI.

Resumen

Objetivo: Documentar la planificación y los procesos metodológicos de un protocolo de revisión de alcance destinado a mapear el conocimiento científico actual sobre el síndrome TEMPI. Introducción: El síndrome TEMPI es una enfermedad rara, adquirida y multisistémica caracterizada por telangiectasias, niveles elevados de eritropoyetina y eritrocitosis, gammapatía monoclonal, colecciones de líquido perinéfrico y cortocircuito intrapulmonar. Justificación: Debido a su rareza, planteamos la hipótesis de que la falta de conocimiento sobre este síndrome puede llevar a un diagnóstico erróneo o retrasado. Una revisión sistemática de la literatura puede contribuir a aumentar el conocimiento y la difusión de información sobre el tema. Metodología: El protocolo fue registrado en el Open Science Framework y estructurado siguiendo el marco metodológico de cinco etapas de Arksey & O'Malley (2005), de acuerdo con las directrices del Instituto Joanna Briggs. La revisión se guía por la pregunta: "¿Cómo puede la evidencia científica disponible contribuir a mejorar el diagnóstico y la atención integral de los pacientes con Síndrome TEMPI?" Se realizará una búsqueda sistemática en el Sistema de Análisis y Recuperación de Literatura Médica (MEDLINE) y en la Biblioteca Virtual em Saúde (BVS), sin restricciones de tiempo. La literatura gris se recuperará mediante Google Scholar. Dos revisores independientes evaluarán la elegibilidad de los estudios. Resultados: Los hallazgos se informarán de acuerdo con PRISMA-ScR (Elementos de informes preferidos para revisiones sistemáticas y extensión de metanálisis para revisiones de alcance) y se presentarán en formatos tabulares y narrativos alineados con los objetivos del estudio y la pregunta de investigación.

Palabras clave: Discrasia de células plasmáticas; Eritrocitosis; Gammapatías monoclonales; Protocolo de Investigación; Síndrome TEMPI.

1. Introduction

TEMPI syndrome is an acquired multisystem disease that manifests preferentially in the fourth or fifth decade of life. It was first documented in 2011(Sykes DB et al., 2011) and the acronym TEMPI defines the characteristics of the syndrome (Schroyens et al., 2012):

(1) T - Telangiectasias - persistence of dilated capillary vessels, forming small punctate red stains or having a spider-like appearance (Rivitti, E A 2024). In TEMPI Syndrome, telangiectasias are most prominent on the face and trunk, and are rarely seen on the lower limbs (Sykes et al., 2011). Although subjectively they appear to be limited to the skin, some patients may have telangiectasias in deep organs such as the ascending colon, cranial bones and dorsal vertebral bodies. Superficial telangiectasias are generally benign, but those in deep organs can lead to serious hemorrhagic events with high morbidity and mortality (Xu et al., 2022)**;**

(2) E - Elevated erythropoietin and erythrocytosis. Erythropoetin is the main regulator of erythropoiesis, which acts by stimulating mitosis and cell differentiation of red blood cell progenitor cells (Anon n.d.). High levels of serum erythropoetin were observed in all patients diagnosed with TEMPI syndrome (Sykes, O'Connell, and Schroyens 2020). Studies suggest that increased erythropoietin stimulates erythrocytosis and angiogenesis, which can aggravate telangiectasias and/or promote increased intrapulmonary shunt with consequent hypoxia which, in turn, can lead to a further rise in erythropoietin (Xu et al., 2022);

(3) M - Monoclonal gammopathy. A condition characterized by the presence of a population of immunoglobulin-producing cells (Faria and Silva 2007). Monoclonal gammopathy is the most striking feature of TEMPI syndrome and possibly plays a crucial role in the pathophysiology of the disease. Among the many monoclonal proteins reported, IgG kappa is the most prevalent (Xu et al., 2022);

(4) P - Peri-nephrotic fluid collections. Fluid accumulations near the kidneys have appeared unilaterally or bilaterally in patients with the syndrome (Máximo 2003). The increase in collections can lead to abdominal fullness, flank pain, nausea, hypertension or the appearance of a palpable abdominal mass (Xu et al., 2022);

(5) I - Intrapulmonary shunt. Intrapulmonary shunt is a manifestation of the syndrome and is accompanied by a decrease in oxygen saturation at rest and, as it progresses, a dependence on supplementary oxygen (Xu et al., 2022).

TEMPI syndrome can be considered an "ultra-rare" disease, a subcategory introduced by the National Institute for Clinical Excellence for diseases with a prevalence of less than <1 per 50000 people (Harari, 2016; Smith, Bergman & Hagey

2022). Although its clinical characteristics are well documented, the etiology, pathophysiology and treatment of TEMPI syndrome remain under investigation. Given its rarity, we postulate that the lack of knowledge about TEMPI syndrome can lead to a misdiagnosis and/or late diagnosis of the disease and that a literature review can contribute to increasing and spreading awareness on the subject.

The scoping review was the methodology chosen because it aims to synthesize knowledge, following a systematic approach to mapping evidence by identifying the main concepts, theories and knowledge gaps (Mattos, Cestari & Moreira 2023).

A preliminary search was carried out on Medline, the Cochrane Database of Systematic Reviews and JBI Evidence Synthesis, and no current or ongoing systematic or scoping reviews on the topic were identified.

According to the methodological guidelines of the Joanna Briggs Institute (JBI), the process of conducting a scoping review begins with the formulation of a review protocol. A scoping review protocol should provide a comprehensive plan for the conduct and reporting of the subsequent scoping review report. The construction of this protocol was carried out before the scope review began, aiming to reduce biases in selection, retrieval and screening of references, ensuring technical rigor and allowing reproducibility (Peters et al., 2020; Tricco et al., 2018). The protocol is a valuable manuscript to publish or make publicly available because it informs the research community of the intentions and focus of the review authors prior to the conduct of the review itself (Peters et al., 2022).

Therefore, to provide a comprehensive overview of TEMPI syndrome, this article aims to present the development of a scoping review protocol, detailing the planning process and methodological approach. The review is designed to map, identify, and summarize the existing scientific evidence on this condition, covering key aspects such as clinical diagnosis, treatment, and pathophysiology.

2. Methodology

This scoping review will be conducted following the JBI methodology for scoping reviews (Peters et al., 2020). The reporting process will adhere to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses extension for scoping reviews (PRISMA-ScR) (Tricco et al., 2018). The review protocol will be aligned with best practices and standardized reporting guidelines, incorporating adapted PRISMA-P (Peters et al., 2022) items helping in the consistency of results and guaranteeing the reproducibility of the scientific study.

This protocol of scoping review is organized in six methodological steps: Stage 1: identifying the research question Stage 2: identifying relevant studies Stage 3: study selection Stage 4: charting the data Stage 5: collating, summarizing and reporting the results.(Arksey & O'Malley, 2005). This scoping review protocol was registered in the Open Science Framework (doi: 10.17605/OSF.IO/TJSD).

Stage 1 - Identifying the research question

The research question of this study is: "How can available scientific evidence contribute to improving the diagnosis and comprehensive care of patients with TEMPI syndrome?" The question was structured by the acronym PCC, where: P (Population) - patients with TEMPI syndrome; C (Concept) diagnosis and comprehensive care (pathophysiology, clinical manifestations and therapy); and C (Context) current knowledge in scientific literature.

The research objectives

1. Map the available scientific evidence on the diagnosis and comprehensive care of patients with TEMPI syndrome, including its pathophysiological, clinical and therapeutic aspects.
2. Identify gaps in scientific knowledge about TEMPI syndrome that can direct future research.

Stage 2 - Identifying relevant studies

The search strategy will aim to locate and include all studies that meet the selection criteria. As TEMPI syndrome is an extremely rare condition and does not have specific indexed terms in the Medical Subject Headings (MeSH) and Health Sciences Descriptors (DeCS), the search strategy was expanded to include not only the term “TEMPI”, but also the main characteristic components of the syndrome. These clinical components were incorporated to increase the sensitivity of the search and ensure that related studies are identified The following are the main components included in the strategy:

- **Telangiectasia** (telangiectasis)
- **Erythrocytosis, polycythemia** (erythrocytosis, polycythemia)
- **Monoclonal gammopathy, Paraproteinemias** (monoclonal gammopathy, monoclonal protein)

Given the scarcity of indexed studies with descriptors directly related to its pathophysiological, clinical and therapeutic aspects, we opted for a search strategy focused mainly on the 'Population' element (patients with TEMPI Syndrome). The intention is to increase the sensitivity of the search, avoiding restricting the results too much and thus guaranteeing the identification of all relevant studies that may address, even indirectly, the topics of interest. The inclusion of additional descriptors for 'Concept' and 'Context' could result in the exclusion of potentially valuable studies due to the absence of specific indexers for the syndrome. For each item of the strategy, a set of descriptors available in Medical Subject Headings (MeSH) and Descritores de Ciências da Saúde (DeCS) as described in Chart 1.

Chart 1 - Terms used in the search strategy.

DeCS terms	**DeCS related terms**
Sindrome TEMPI	TEMPI
TEMPI Syndrome	TEMPI disorder
Telangiectasia	
Policitemia	Eritrocitose
Polycythemia	Erythrocytoses, Erythrocytosis
Paraproteinemias	Gamopatia Monoclonal
	Discrasia de plasmocitos
Paraproteinemias	Monoclonal gammopathy

MeSH terms	**MeSH related terms**
TEMPI Syndrome	
Telangiectasis	
Polycythemia	Erythrocytoses, Erythrocytosis
Paraproteinemias	Monoclonal gammopathy

Source: Authors (2024).

Inclusion Criteria: The definition of eligibility criteria will follow the acronym PCC to answer the review question, where**:** **population:** Studies on patients diagnosed with TEMPI syndrome, regardless of age group, ethnicity, sex or comorbidities; **concept:** what are the most effective approaches, based on available scientific evidence, to improve the TEMPI syndrome diagnosis process, as well as to develop personalized care strategies that address patients' individual needs and improve their quality of life. How healthcare professionals can improve the identification and management of this condition, taking into account the diagnostic challenges and specific needs of patients. What interventions or care are aimed at early diagnosis and clinical management for TEMPI syndrome; **context:** the current scientific knowledge on the topic.

types of evidence sources: This scoping review will consider articles published in any language, in the public and private domain, and with different methodological approaches, without time or language restrictions in the search for evidence. Texts

and opinion articles will also be considered for inclusion in this scoping review. If more information is needed, the author of the primary study can be contacted.

Exclusion Criteria Duplicate studies, or studies whose full text is not available, or documents that do not meet the inclusion criteria will be excluded.

The search strategy will be aimed at recruiting published and unpublished studies, using combinations of descriptors and keywords. The Medical Literature Analysis and Retrieval System (MEDLINE) via PubMed, the Biblioteca Virtual em Saude (BVS) and the Scientific Electronic Library Online (SciELO) will be used as verified data sources for this review. The search for gray literature will be carried out using Google Scholar. Chart 2 specifies the search strategy in Medline (via Pubmed), which will later be adapted for other databases. During the development of the review, if the reviewers identify any keywords, free terms or additional sources of interest, these will be incorporated into the search strategy and reported transparently in the final version of the review.

Chart 2 - Search strategy in Medline (via Pubmed).

Search number	Search	Keywords	Results
6	(#1) OR (#5)	"TEMPI Syndrome"[All Fields] OR (("telangiectasic"[All Fields] OR "telangiectasis"[MeSH Terms] OR "telangiectasis"[All Fields] OR "telangiectases"[All Fields]) AND ("polycythaemia"[All Fields] OR "polycythemia"[MeSH Terms] OR "polycythemia"[All Fields] OR "polycythaemias"[All Fields] OR "polycythemias"[All Fields] OR ("polycythemia"[MeSH Terms] OR "polycythemia"[All Fields] OR "erythrocytosis"[All Fields]) OR ("polycythemia"[MeSH Terms] OR "polycythemia"[All Fields] OR "erythrocytosis"[All Fields])) AND ("paraproteinaemia"[All Fields] OR "paraproteinemias"[MeSH Terms] OR "paraproteinemias"[All Fields] OR "paraproteinemia"[All Fields] OR "paraproteinaemias"[All Fields] OR ("paraproteinemias"[MeSH Terms] OR "paraproteinemias"[All Fields] OR ("monoclonal"[All Fields] AND "gammopathy"[All Fields]) OR "monoclonal gammopathy"[All Fields] OR "monoclonal gammopathy of undetermined significance"[MeSH Terms] OR ("monoclonal"[All Fields] AND "gammopathy"[All Fields] AND "undetermined"[All Fields] AND "significance"[All Fields]) OR "monoclonal gammopathy of undetermined significance"[All Fields]))	42
5	((#2) AND (#3)) AND (#4)	("telangiectasic"[All Fields] OR "telangiectasis"[MeSH Terms] OR "telangiectasis"[All Fields] OR "telangiectases"[All Fields]) AND ("polycythaemia"[All Fields] OR "polycythemia"[MeSH Terms] OR "polycythemia"[All Fields] OR "polycythaemias"[All Fields] OR "polycythemias"[All Fields] OR ("polycythemia"[MeSH Terms] OR "polycythemia"[All Fields] OR "erythrocytosis"[All Fields]) OR ("polycythemia"[MeSH Terms] OR "polycythemia"[All Fields] OR "erythrocytosis"[All Fields])) AND ("paraproteinaemia"[All Fields] OR "paraproteinemias"[MeSH Terms] OR "paraproteinemias"[All Fields] OR "paraproteinemia"[All Fields] OR "paraproteinaemias"[All Fields] OR ("paraproteinemias"[MeSH Terms] OR "paraproteinemias"[All Fields] OR ("monoclonal"[All Fields] AND "gammopathy"[All Fields]) OR "monoclonal gammopathy"[All Fields] OR "monoclonal gammopathy of undetermined significance"[MeSH Terms] OR ("monoclonal"[All Fields] AND "gammopathy"[All Fields] AND "undetermined"[All Fields] AND "significance"[All Fields]) OR "monoclonal gammopathy of undetermined significance"[All Fields]))	23
4	(paraproteinemia) OR (Monoclonal gammopathy)	"paraproteinaemia"[All Fields] OR "paraproteinemias"[MeSH Terms] OR "paraproteinemias"[All Fields] OR "paraproteinemia"[All Fields] OR "paraproteinaemias"[All Fields] OR ("paraproteinemias"[MeSH Terms] OR "paraproteinemias"[All Fields] OR ("monoclonal"[All Fields] AND "gammopathy"[All Fields]) OR "monoclonal gammopathy"[All Fields] OR "monoclonal gammopathy of undetermined significance"[MeSH Terms] OR ("monoclonal"[All Fields] AND "gammopathy"[All Fields] AND "undetermined"[All Fields] AND "significance"[All Fields]) OR "monoclonal gammopathy of undetermined significance"[All Fields])	67,705

3	((polycythemia) OR (Erythrocytosis)) OR (Erythrocytosis)	"polycythaemia"[All Fields] OR "polycythemia"[MeSH Terms] OR "polycythemia"[All Fields] OR "polycythaemias"[All Fields] OR "polycythemias"[All Fields] OR "polycythemia"[MeSH Terms] OR "polycythemia"[All Fields] OR "erythrocytosis"[All Fields] OR "polycythemia"[MeSH Terms] OR "polycythemia"[All Fields] OR "erythrocytosis"[All Fields]	17,333
2	telangiectasis	"telangiectasic"[All Fields] OR "telangiectasis"[MeSH Terms] OR "telangiectasis"[All Fields] OR "telangiectases"[All Fields]	12,664
1	"TEMPI Syndrome"	"TEMPI Syndrome"[All Fields]	40

Source: Authors (2024).

After searching the databases, the results will be exported to Rayyan® Software (Qatar Computing Research Institute) (Ouzzani et al., 2016), where they will be grouped to remove duplicate articles. The results of the search and study inclusion process will be reported in full in the final scoping review, presented in the form of a flowchart, detailing the results of the sources of evidence (databases and additional sources), removal of duplicates, screening and selection of studies in phase 1 and 2, with the reasons why they were excluded, and the number of studies included in the review according to the PRISMA-ScR flowchart (Mattos et al., 2023).

Stage 3- Study selection

The identification of relevant studies will be based on the eligibility criteria guided and directed by the research question. Initially, a pilot test of the selection of articles will be carried out to adjust the criteria among the researchers. Thereafter, the titles and abstracts will be examined independently by the two researchers (phase 1). The reasons for excluding sources of full-text evidence that do not meet the inclusion criteria will be recorded and reported. Any disagreements that arise between the reviewers will subsequently be resolved through discussion or, if necessary, with the participation of a third reviewer. Next, the potentially relevant sources will be retrieved in full and the full text of the articles screened will be read (phase 2) to determine the final inclusion of the studies according to the inclusion criteria previously explained.

Stage 4 - Charting the data

This scoping review is designed to map the identified data, without the act of synthesis or reference to methodological quality of the studies. Data extraction from the studies included in the review will be carried out independently by the two reviewers, using a form (Chart 3) developed by the authors adapted from the JBI data extraction tool (Peters et al., 2020). The data extracted will include specific details about the participants, the concept, the context, the study methods and the main results relevant to the review questions. A pilot test will be carried out on as many publications as necessary to familiarize the reviewers with the extraction tool. Any differences between the reviewers' selections will be resolved by consensus or decided jointly with a third reviewer.

Chart 3 - Data extraction form.

Information	**Description**
	Bibliographic information
Authors	Authors' surname and first name
Year	Year of publication
Journal	Journals' name
Title	Original title of the publication
Level of evidence	Level of evidence JBI

Details extracted from source of evidence	
Objective	Describe the main objective of the study
Participant characteristics	Describe the main characteristic of the participants
TEMPI diagnostic criteria	Describe the TEMPI criteria
Laboratory	Describe the main laboratory findings
Treatment	Describe the treatment used
Pathophysiology	Describe the pathophysiology discussed in the study
Contribution	Describe the main contributions of the study
Final considerations	Describe the main conclusions of the study

Source: Authors (2024).

The instrument is preliminary and may be updated, incorporating any relevant data identified during the review. Any changes will be clearly described and justified in the final version of the review.

Stage 5 - Collating, summarizing and reporting the results.

In this scoping review, the extracted data will be presented in tables, charts, and/or images, following the PRISMA-ScR guidelines (Tricco et al., 2018) and aligned with the review's objective. The data will be categorized to facilitate mapping and knowledge generation, with evidence synthesized into a narrative format. A thematic analysis will be conducted to identify key themes emerging from the extracted data, which will then be described narratively to summarize the study findings

3. Final Considerations

This protocol outlines the methodological steps for conducting a scoping review on TEMPI Syndrome, aiming to clarify the process and reduce biases in literature search and article selection. Our goal is to publish the findings in respected academic medical journals and present them at relevant scientific conferences, contributing to the expansion of knowledge on TEMPI syndrome. Ultimately, we hope this will support healthcare professionals in diagnosing this rare multisystem disease early, enhancing patient prognosis through better clinical awareness.

Acknowledgments

We would like to thank the Universidade Municipal de São Caetano do Sul - USCS for providing the fertile ground for research and Dr. David Sykes, as a pioneer in the description of the disease and a source of consultations on the subject.

References

Arksey, H., & O'Malley, L. (2005). Scoping studies: towards a methodological framework. *International Journal of Social Research Methodology*, 8, 19-32. 10.1080/1364557032000119616

Faria, Rosa Malena D., and Roberta O. Paula e Silva. 2007. “Gamopatias monoclonais: critérios diagnósticos e diagnósticos diferenciais.” *Revista Brasileira de Hematologia e Hemoterapia* 29:17–22. doi: https://doi.org/10.1590/S1516-84842007000100005.

Harari, Sergio. 2016. “Why We Should Care about Ultra-Rare Disease.” *European Respiratory Review* 25(140):101–3. doi: 10.1183/16000617.0017-2016.

Mattos, Samuel Miranda, Virna Ribeiro Feitosa Cestari, and Thereza Maria Magalhães Moreira. 2023. “Scoping protocol review: PRISMA-ScR guide refinement.” *Rev Enferm UFPI* 12(1). doi: 10.26694/reufpi.v12i1.3062.

Máximo, Carlos Henrique M. 2003. “Acesso percutâneo nas coleções e pseudocistos pancreáticos.” *Radiologia Brasileira* 36:168–168. doi: https://doi.org/10.1590/S0100-39842003000300015.

Ouzzani, Mourad, Hossam Hammady, Zbys Fedorowicz, and Ahmed Elmagarmid. 2016. “Rayyan—a Web and Mobile App for Systematic Reviews.” *Systematic Reviews* 5(1):210. doi: 10.1186/s13643-016-0384-4.

Peters, Micah D. J., Casey Marnie, Andrea C. Tricco, Danielle Pollock, Zachary Munn, Lyndsay Alexander, Patricia McInerney, Christina M. Godfrey, and

Hanan Khalil. 2020. “Updated Methodological Guidance for the Conduct of Scoping Reviews.” *JBI Evidence Synthesis* 18(10):2119–26. doi: 10.11124/JBIES-20-00167.

Peters, Micah D.J.1,2,3; Godfrey, Christina; McInerney, Patricia; Khalil, Hanan; Larsen, Palle8; Marnie, Casey; Pollock, Danielle; Tricco, Andrea C.; Munn, Zachary. Best practice guidance and reporting items for the development of scoping review protocols. JBI Evidence Synthesis 20(4):p 953-968, April 2022. | DOI: 10.11124/JBIES-21-00242

Rivitti, E A. 2024. *Manual de Dermatologia Clinica de Sampaio e Rivitti - Rivitti. - Livraria Florence*. (2nd ed.). Grupo A.

Schroyens, A., B. Sykes, L. Kwok, C. Landgren, and L. O’Connell. 2012. Description of a rare and novel disease: tempi syndrome. 97, 608–608.

Smith, C. I. Edvard, Peter Bergman, and Daniel W. Hagey. 2022. “Estimating the Number of Diseases – the Concept of Rare, Ultra-Rare, and Hyper-Rare.” *iScience* 25(8), 104698. doi: 10.1016/j.isci.2022.104698.

Sykes, David B., Casey O’Connell, and Wilfried Schroyens. 2020. “The TEMPI Syndrome.” *Blood* 135(15), 1199–1203. doi: 10.1182/blood.2019004216.

Sykes DB, Schroyens W, and O’Connell C. 2011. The TEMPI Syndrome--a Novel Multisystem Disease. *The New England Journal of Medicine* 365(5), 475–77. doi: 10.1056/NEJMc1106670.

Tricco, Andrea C., Erin Lillie, Wasifa Zarin, Kelly K. O’Brien, Heather Colquhoun, Danielle Levac, David Moher, Micah D. J. Peters, Tanya Horsley, Laura Weeks, Susanne Hempel, Elie A. Akl, Christine Chang, Jessie McGowan, Lesley Stewart, Lisa Hartling, Adrian Aldcroft, Michael G. Wilson, Chantelle Garritty, Simon Lewin, Christina M. Godfrey, Marilyn T. Macdonald, Etienne V. Langlois, Karla Soares-Weiser, Jo Moriarty, Tammy Clifford, Özge Tunçalp, and Sharon E. Straus. 2018. “PRISMA Extension for Scoping Reviews (PRISMA-ScR): Checklist and Explanation.” *Annals of Internal Medicine* 169(7), 467–73. doi: 10.7326/M18-0850.

Xu, Jian, Wenqi Liu, Fengjuan Fan, Bo Zhang, Fei Zhao, Yu Hu, and Chunyan Sun. 2022. “TEMPI Syndrome: Update on Clinical Features, Management, and Pathogenesis.” *Frontiers in Endocrinology* 13, 886961. doi: 10.3389/fendo.2022.886961.