MARIA LUIZA CIMARDI RUPP

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Trabalho apresentado à Universidade Federal de Santa Catarina, como requisito para a conclusão do Curso de Graduação em Medicina.

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Presidente do Colegiado: Profº. Dr. Edevard José de Araújo
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Este trabalho é dedicado aos meus avós, aos meus pais e a todos aqueles que me apoiaram nos últimos anos. É também dedicado aos pequenos que me dão mais força do que imaginam: Felipe, Pietro e todas as crianças com as quais tenho o prazer de conviver.

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Agradeço aos meus pais, que nunca mediram esforços para me dar o suporte necessário em todos os projetos que tive ao longo da vida.

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Mucinose folicular na infância: um diagnóstico raro, mas significante

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Título curto: Mucinose folicular e seu significante diagnóstico

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Abreviações:
MF: Follicular mucinosis (Mucinose folicular)
MFg: Mycosis fungoides
LSC: Lichen simplex chronicus.

Resumo do conteúdo: Este Relato de Caso ressalta a importância de pediatras generalistas saberem diagnosticar a Mucinose Folicular e seu impacto no prognóstico dos pacientes.

Página de declaração dos contribuidores

Maria Luiza Cimardi Rupp desenhou o estudo, realizou a análise de dados, redigiu o manuscrito original, realizou a revisão de literatura e revisou o manuscrito.

Marice El Achkar Mello desenhou o estudo, realizou a coleta e a análise inicial de dados e revisou e corrigiu o manuscrito.

Maria Marlene de Souza Pires desenhou o estudo, coordenou e supervisionou a coleta de dados e revisou e corrigiu o manuscrito.

Amanda Amaro Pereira realizou a análise de dados e revisou e corrigiu o manuscrito.

Todos os autores aprovaram o manuscrito final como submetido e concordaram em ser responsáveis por todos os aspectos do trabalho.

Abstract:
Follicular mucinosis (MF) is a rare cutaneous disease with unclear etiology, it was firstly described by Pinkus et. al. in 1957, who named it mucinous alopecia, referring to a characteristic process of the MF, which leads to alopecia. As this process was not seen on all developments of the disease, in 1959 Jablonska et. al. proposed to name it Follicular mucinosis. Follicular mucinosis is uncommon in children. Using the online searching databases Scielo and PubMed, during the past 10 years, there were found only 16 pediatric case reports about the disease: 3 associated with mycosis fungoides (MFg), 1 about the acneiform presentation of the disease, 9 discussing the primary type, 1 did not specify the diagnosis in the report and 2 were about other presentations of the disease. The main concern of diagnosing a child with MF is whether it is associated with MFg, a cutaneous T-cell lymphoma, that although rare in children, when present is mostly associated with MF. The earlier the diagnosis of MFg is made, the better is the prognosis of the patient. Therefore, it is of great importance that a general practitioner pediatrician can diagnose a case of MF and identify whether it is associated with MFg or other systemic diseases.

Resumo:
Mucinose folicular (MF) é uma doença cutânea rara e de etiologia incerta, mas que pode impactar drasticamente a qualidade de vida e o prognóstico dos pacientes. Dessa forma, o presente estudo busca divulgar, no meio científico, uma das possíveis apresentações clínicas da doença, além de reforçar a importância do seu diagnóstico e acompanhamento. A MF foi descrita pela primeira vez por Pinkus et. al. em 1957, que a nomeou Alopecia mucinosa referindo-se a um processo característico da MF, que leva a alopecia. Entretanto, como esse processo não ocorre em todos os casos da doença, em 1959, Jablonska et. al. propôs a alteração do nome para mucinose folicular. A MF é incomum em crianças: usando as bases de dados online da Scielo e do PubMed, foram encontrados apenas 16 relatos de casos pediátricos da doença durante os últimos 10 anos. Entre esses relatos, 3 eram associados à micose fungoide (MFg), 1 sobre a apresentação acneiforme (primária) da doença, 9 discutindo a forma primária clássica da doença, 1 não especificou o diagnóstico no relato e 2 são sobre outras apresentações da doença. A principal preocupação ao fazer um diagnóstico de mucinose folicular é investigar se ele é ou não associado à micose fungoide. A MFg é um linfoma cutâneo de células T, cujo diagnóstico precoce é essencial para um melhor prognóstico do paciente. Apesar de rara em crianças, quando a MFg está presente, é comumente associada a MF. Dessa forma, é de grande importância que um médico generalista consiga identificar um caso de mucinose folicular a fim de dar o encaminhamento necessário ao paciente, excluindo possíveis malignidades associadas.

Introdução

Mucinose folicular (MF) é uma doença cutânea rara com etiologia pouco esclarecida[1]. MF pode ocorrer como uma desordem primária (idiopática) ou secundária a doenças benignas ou malignas, como micose fungoide (MFg), Lúpus[2] ou outras doenças de pele, como o impetigo. A apresentação clínica da doença é variável e os principais achados incluem: presença de placas ou pápulas bem delimitadas, eritematosas ou acastanhado-eritematosas, que aparecem de maneira aguda ou subaguda, principalmente na cabeça e/ou pescoço[2]. Manifestações menos usuais incluem lesões similares a alopecia cicatricial, alopecia areata, cistos, nódulos, eczema e erupções acneiformes crônicas[3]. MF é caracterizada histopatologicamente pela deposição de mucina nas glândulas sebáceas e no epitélio folicular. Por causa de sua etiologia incerta, o exame histopatológico é necessário para confirmar o diagnóstico.

Segundo as bases de dados online da Scielo e do PubMed, foram encontrados apenas 16 relatos de caso da doença durante os últimos 10 anos. Entre esses relatos, 3 estão associados à micose fungoide (MFg), 1 à apresentação acneiforme (primária) da doença, 9 à discussão da forma primária clássica da doença, 1 que não especificou o diagnóstico no relato e 2 sobre outras apresentações da doença. Entre os casos relatados, é importante destacar, como principais diagnósticos diferenciais, o líquen espinuloso (Fig. 1) e a ceratose pilar (Fig. 2), ambos são os principais diagnósticos diferenciais para crianças com lesões suspeitas. As lesões geralmente têm bom prognóstico e se resolvem de maneira espontânea, especialmente em crianças mais novas[1-6,] entretanto, algumas lesões podem ser secundárias a linfomas cutâneos de células-T, as quais requerem tratamento mais intensivo, além de acompanhamento clínico em longo prazo[2-5, 7-12].

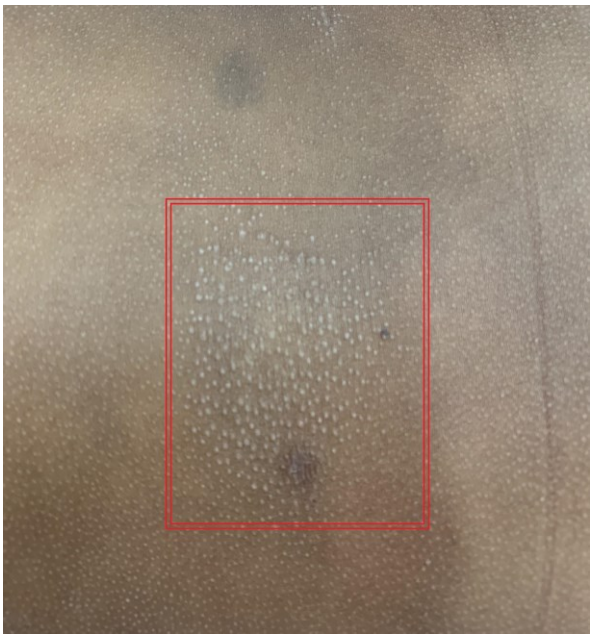

Figura 1 - (Arquivo pessoal Marice El Achkar Mello, MD)

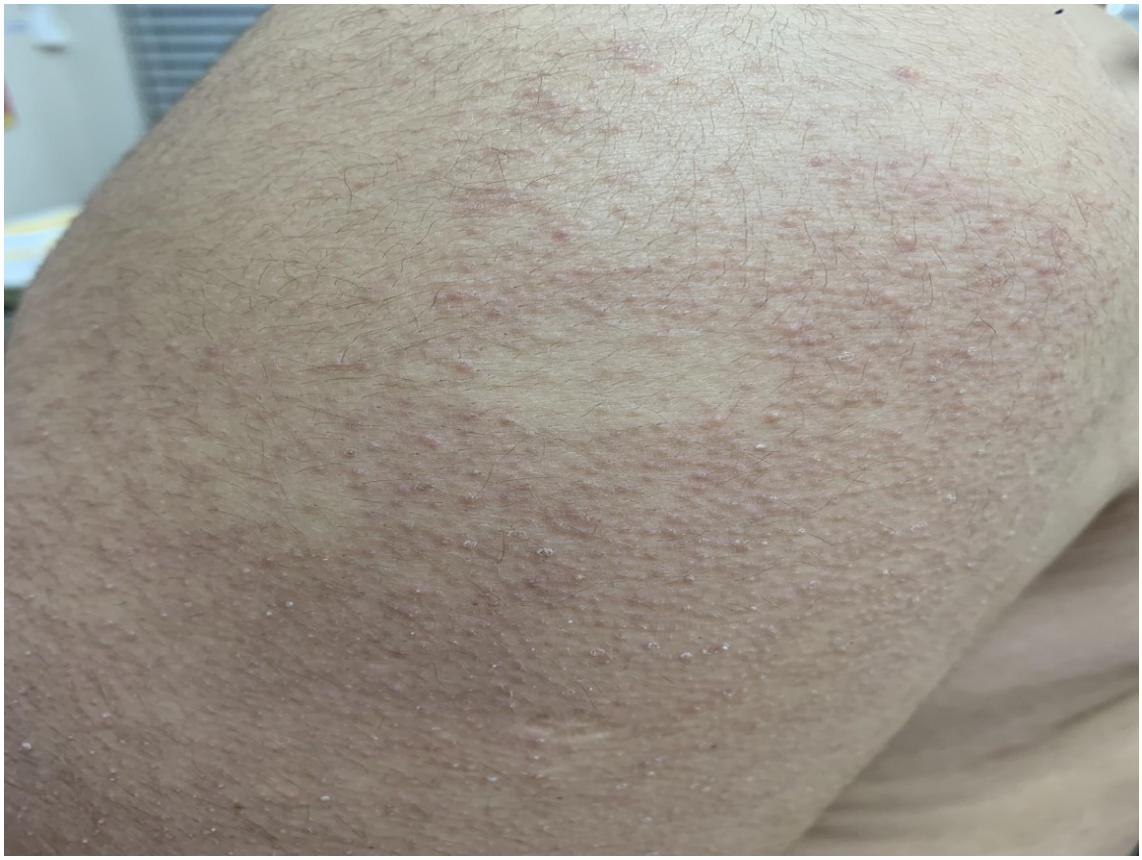

Figura 2 - (Arquivo pessoal Marice El Achkar Mello, MD)

Apresentação do Caso

Paciente masculino, de 5 anos de idade, apresentava pápulas eritematosas, que apareceram cerca de 1 ano antes da consulta, distribuídas linearmente ao longo de 5 cm na parte posterior da panturrilha esquerda (Fig. 3.a). Ele reclamava de prurido no local e a lesão estava escoriada. No início do quadro, a criança havia sido tratada por outro profissional para impetigo e larva migrans cutânea, usando antibióticos e antifúngicos tópicos, sem resposta aos tratamentos. A próxima hipótese clínica foi a de líquen simples crônico, haja vista que o quadro clínico permanecia o mesmo e não houve resposta aos tratamentos prévios. Assim, a próxima linha de tratamento foi o uso de corticosteroide tópico oclusivo por 30 dias, que apresentou redução do prurido e das lesões.

Quatro meses após o fim do tratamento, as lesões retornaram com a mesma apresentação clínica de pápulas eritematosas acompanhadas de prurido (Fig. 3.b). Foi feita biópsia da lesão e, após avaliação histológica (Fig.4), diagnosticada a mucinose folicular. Os achados histopatológicos mostraram siringometaplasia e infiltrado linfoide associados. Essas condições poderiam corresponder à entidade primária, ou secundária, reacional, dada a história de impetigo prévio. A primeira linha de tratamento utilizada foi um inibidor de calcineurina por 60 dias, sem resposta ao tratamento. Posteriormente, foi reiniciado o tratamento com corticosteroide tópico oclusivo por 60 dias, com boa resposta ao tratamento e sem recidivas até o momento. Apesar da ausência de recidivas, não é possível diferenciar se o caso foi idiopático ou secundário ao impetigo relatado pelo médico anterior. Considerando a idade e o quadro clínico do paciente, é improvável que haja outras doenças malignas associadas, como a micose fungoide, apesar disso, o paciente foi orientado a retornar ao serviço médico em caso de recidivas.

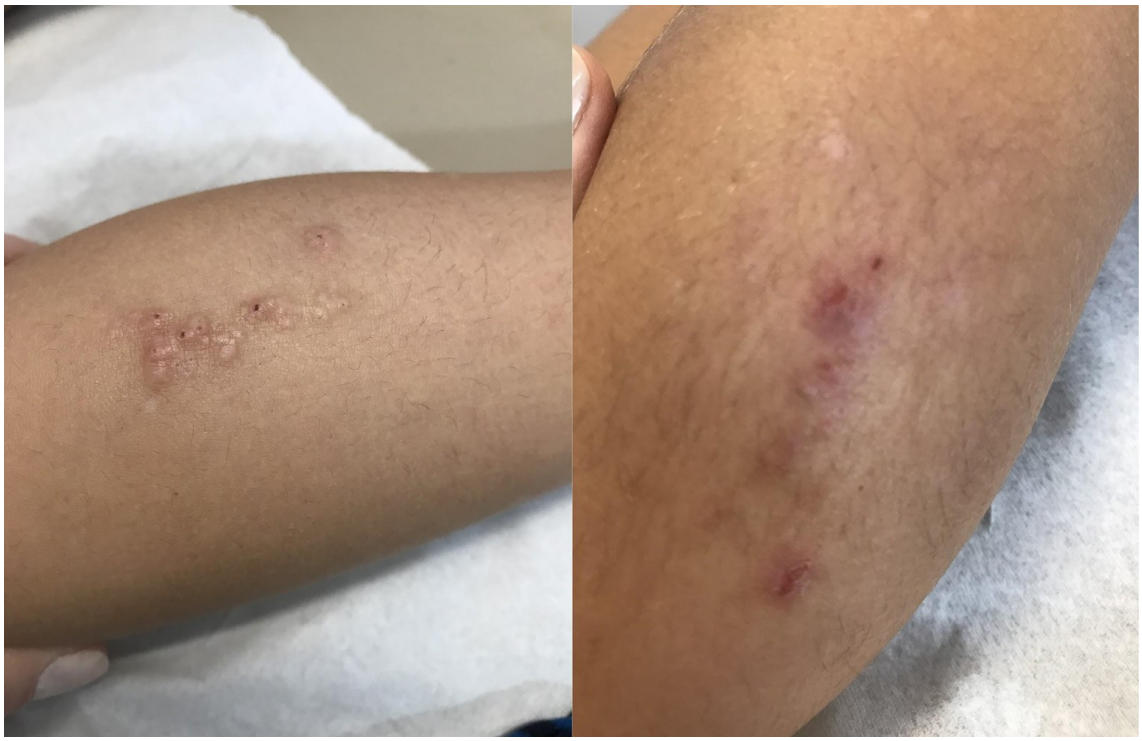

Figura 3.a - Figura 3.b - (Arquivo pessoal Marice El Achkar Mello, MD)

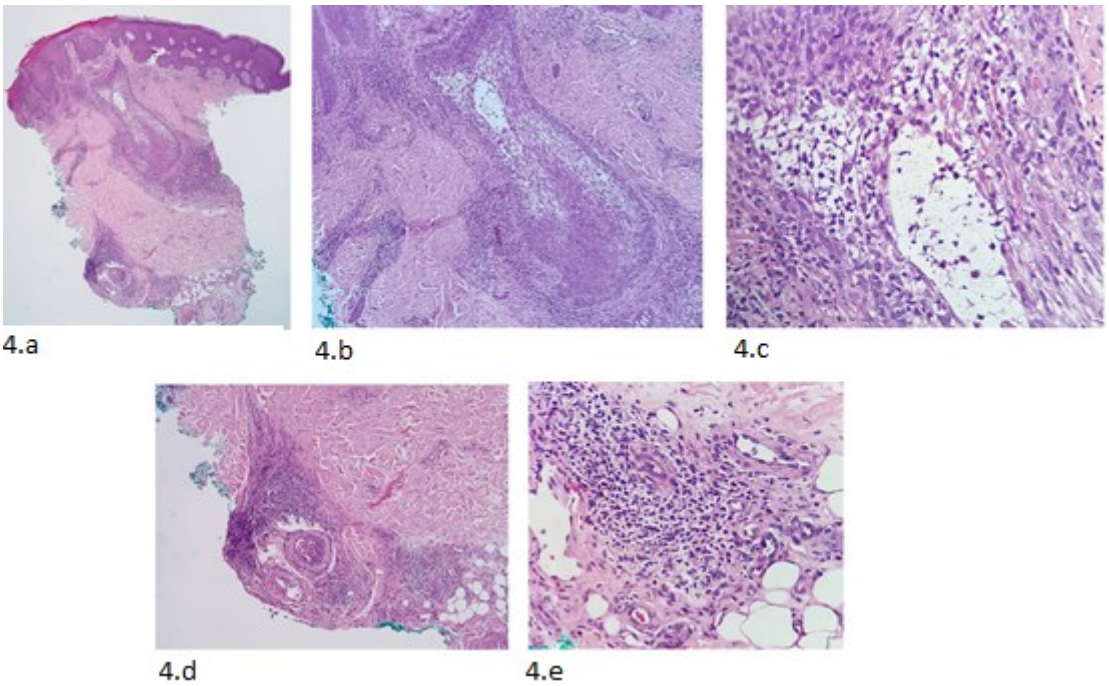

Figura - (Arquivo pessoal Marice El Achkar Mello, MD e Amanda Amaro Pereira, MD)

Discussão

Mucinose folicular foi descrita pela primeira vez em 1957, por Pinkus et.al. usando o termo "Alopecia Mucinosa"[1, 5, 8-10, 13], sendo mais tarde descrita como mucinose folicular por Jablonska et.al. em 1959[1, 5, 8, 13]. A etiologia da doença permanece incerta, porém, foram descritas quatro manifestações clínicas principais, três delas descritas por Coskey e Mehregan em 1970[10].

A primeira manifestação, conhecida como mucinose folicular benigna, é primária e tem uma evolução curta, geralmente afetando crianças e adultos jovens. A segunda, conhecida como MF associada a linfoma, também é uma desordem primária e compreende condições como a micose fungoide, sendo mais comum em adultos mais velhos. A terceira forma da MF é secundária a doenças malignas e afeta principalmente pacientes idosos. Por fim, a quarta manifestação é conhecida como mucinose folicular reativa, referindo-se aos achados histopatológicos que descrevem a MF, mas são encontrados em uma variedade de neoplasias e dermatoses.[10]

Mucinose folicular é rara em crianças[8] e os principais diagnósticos diferenciais são o líquen espinuloso e a ceratose pilar. O Diagnóstico da MF é confirmado apenas após biópsia e avaliação histológica da pele afetada, a qual deve demonstrar: acúmulo de mucina no infundíbulo folicular e glândula sebácea adjacente, com separação dos queratinócitos; graus variáveis de inflamação perifolicular também podem ser observados, com predominância de linfócitos e eosinófilos. Em casos raros, pode haver presença de siringometaplasia, como no paciente relatado neste trabalho. Quando a MF é associada a linfoma cutâneo, pode haver foliculotropismo mais evidente, o qual pode ou não ser associado à epidermotropismo e à atipia celular.

Um algoritmo de tratamento ainda não foi estabelecido, mas pode incluir medicamentos antimaláricos, corticoides sistêmicos ou tópicos, além de outras medicações[8]. O tratamento utilizado no caso apresentado foi um corticosteroide oclusivo tópico de média potência. A escolha da medicação foi baseada em referências consistentes da literatura, de consenso dentro da comunidade médica, além do fácil acesso a ela e seu baixo custo, ou seja, melhor relação custo, risco/benefício em comparação com as alternativas terapêuticas disponíveis.

O paciente teve boa resposta ao tratamento, apesar disso, avaliações clínicas em longo prazo, com biópsia das lesões remanescentes, são importantes para excluir associação com MFg. A micose fungoide (MFg) é um linfoma de célula T cutâneo e, como acontece com a Mucinose Folicular, não possui algoritmo de tratamento para casos pediátricos. Algumas de suas apresentações clínicas tendem a indicar bom prognóstico, como lesões únicas, localizadas na cabeça ou no pescoço e que foram diagnosticadas ainda na infância.

É importante ressaltar que, apesar de rara em crianças e adultos jovens, a micose fungoide requer diagnóstico precoce para garantir um bom prognóstico. Dessa forma, é crucial que pediatras saibam identificar e diagnosticar lesões causadas pela mucinose folicular a fim de avaliar se sua manifestação é primária, associada à Micose fungoide, ou secundária a outras doenças. Em casos de MF secundária, o tratamento deve ser direcionado ao controle da doença de base, como o Lúpus, por exemplo. Mais estudos sobre a relação entre essas duas doenças são necessários, tendo em vista que ainda não há uma descrição clara de sua fisiopatologia, situação que exige avaliações médicas seriadas até resolução completa das lesões.

Conclusão

É importante o diagnóstico precoce da Mucinose Folicular, pois, embora rara, a doença pode ter prognóstico ruim quando associada à micose fungoide ou a outras doenças de base. Assim, a intervenção nesses casos é especialmente relevante para melhora do prognóstico dessas e de outras doenças potencialmente graves que podem estar relacionadas à etiologia da Mucinose folicular.

Além disso, mesmo no caso das formas idiopáticas da Mucinose folicular, há o transtorno causado ao paciente e à família, seja pelo prurido constante seja pelos diversos diagnósticos não resolutivos, já que a doença é rara e pouco conhecida, o que leva, consequentemente, a tratamentos equivocados e à redução da qualidade de vida.

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Figura 1 - Líquen Espinuloso: pápulas esbranquiçadas agrupadas formando uma placa, em região do tronco.

Figura 2 - Ceratose pilar: pápulas cor da pele com base eritematosa em região extensora do membro superior.

Figura 3 – Lesões de pele na primeira consulta (a) e no momento da biópsia – após 4 meses (b).

Figura 4 – Avaliação histológica da biópsia (Hematoxilina-eosina) a, b- Folículo piloso central contendo mucina intercelular; c- Imagem aproximada mostrando infiltrado perifolicular e separação dos queratinócitos; d, e- infiltrado circundando plexos vasculonervosos e glândulas écrinas, com áreas de siringometaplasia.

ANEXO: NORMAS DE PUBLICAÇÃO DA REVISTA

1.1**ACCEPTANCE CRITERIA**

Relevance to readers is of primary importance in manuscript selection. The readership includes general and specialist pediatricians, pediatric researchers and educators, and child health policy-makers. *Pediatrics* receives many more high-quality manuscripts than can be accommodated in our available space. The acceptance rate is approximately 10%. An article that is thought by the editors to not be relevant to readers, outside of scope, or very unlikely to be accepted may be rejected without review. All manuscripts considered for publication are peer reviewed, including those written by members of the Editorial Board. Peer reviewers are selected by the editors. Selection is based on their expertise in the topic of the manuscript. Generally, at least 2 reviews are required before a decision is rendered. Authors can suggest reviewers who they believe should not review the manuscript but should provide a clear rationale for this request.

Authors should carefully follow instructions for manuscript preparation and ensure that the manuscript is proofread before submission. Manuscripts that do not follow the author instructions will not be considered for review. Careless preparation of a manuscript raises concerns about the quality of the work and makes acceptance less likely. Manuscripts are electronically scanned for plagiarism. Authors will be contacted if there is concern about potential plagiarism. *Pediatrics* follows the recommendations of the Committee on Publication Ethics for concerns about plagiarism or any other manuscript-related ethical issue. Manuscripts are judged on the importance, originality, scientific strength, clinical relevance, and clarity of content. *Pediatrics* does not publish manuscripts that focus only on animal research. Refer to the sections below on the particular considerations for each of the manuscript types that appear in the journal. Authors should also consider the comprehensive reporting guidelines for a wide variety of study designs that are available at http://www.equator-network.org/home/. These can be helpful in improving manuscript clarity and completeness. Note that authors submitting manuscripts describing adverse drug or medical device events or product problems should also report these to the appropriate governmental agency. Responses to a published article should be submitted as online comments. The editors will determine which comments will be published in the journal as Letters to the Editor.

After the reviews are received, the editors may take one of the following actions: *Accept*; *Accept with Revisions*; *Reject with option to Resubmit*; *Reject,* or *Reject and Transfer (if authors opted to have their manuscript transferred to* Hospital Pediatrics *in not accepted by* Pediatrics*)*. A rejected manuscript may not be resubmitted. A manuscript may be rejected with an option to resubmit with extensive revision. The resubmitted manuscript receives an additional round of peer review (which may include new reviewers), and the manuscript may or may not be accepted. A decision of *Accept with Revision* indicates that the editors intend to accept the manuscript contingent on adequate response to reviewers. A decision of *Accept*, which is exceedingly rare on first submission, indicates that the manuscript is ready to place into production without further modification. Appeals on decisions will be considered by the editorial board on a case-by-case basis.

1.2**PUBLICATION ETHICS**

Authorship. An “author” is someone who has made substantive intellectual contributions to a published study. Each author is required to meet ALL FOUR of the following criteria:

1. Substantial contribution(s) to conception and design, acquisition of data, or analysis and interpretation of data; **and**
2. Drafting the article or revising it critically for important intellectual content; **and**
3. Final approval of the version to be published, **and**
4. Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

NOTE: Acquisition of funding, collection of data, or general supervision of the research group alone does not constitute a sufficient basis for authorship.

All persons listed as authors must meet these criteria, and all persons who meet these criteria must be listed as authors. Articles submitted with an unexpectedly large number of authors invite scrutiny by editors and reviewers for clear justification for the presence of each person on the authorship list. *Pediatrics* permits a statement of equal contribution for two first authors and two senior authors. On the title page, include asterisks by each name and a statement that reads: * *Contributed equally as co-first authors* or **Contributed equally as co-senior authors.*

Decide authorship issues, including the order, before submission. *Pediatrics* does not allow addition or removal of authors or changes to the author order after a manuscript is submitted without explicit approval from the editors.

If published, author names and affiliations will appear as seen in the submitted manuscript Word document and the final typeset proofs. All authors must ensure that their information is correct.

Conflict of Interest and Disclosure. After a paper is accepted by *Pediatrics* for publication, all authors must submit conflict of interest and disclosure forms. *Pediatrics* adheres to the policy and uses the standardized disclosure form of the International Committee of Medical Journal Editors (ICMJE). The collection of the forms is automated within the online system.

IRB Approval. All studies that involve human subjects must be approved or deemed exempt by an official institutional review board; this should be noted in the Methods section of the manuscript.

Industry Sponsorship. All industry sponsorship must be declared in the manuscript. Manuscripts in which all authors are employed by a commercial entity can raise additional scrutiny from the editorial board.

Registration of Clinical Trials. All clinical trials must be registered in a World Health organization-approved Clinical Trial registry prior to enrollment of the first subject. The registry name and registration number should be included on the title page. Reports of unregistered trials will be returned to authors without review. Publication of the results of a trial that was initiated prior to the ICMJE requirement for trial registration will be considered by the editors on a case-by-case basis.

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Editor Conflict of Interest: Journal editors recuse themselves from manuscripts for which they have a conflict of interest.

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1.4**FORMATTING REQUIREMENTS**

All submissions must adhere to the following format:

- Times New Roman font, size 12, black
- Title Page, Contributors' Statement Page, Abstract, Acknowledgments, and References should be **single-spaced**
- Only the Main Body Text should be **double-spaced**
- Main Submission Document as a Microsoft Word file (no PDFs)
- Include line and page numbering in your Word document (excluding the References).
- Do **not** include page headers or footers in new submissions.
- Do **not** include footnotes within the manuscript body. Footnotes are allowed only in tables/figures.

Refer to the "Article Types" section for specific guidelines on preparing a manuscript in each category. Note in particular the requirements regarding abstracts for different categories of article.

1.5**TITLE PAGE**

The Title Page should appear first in your manuscript document if selecting single-blind peer review, or as a separate file if selecting double-blind peer review. If you select double-blind peer review and are including acknowledgments, those should appear at the end of the Title Page file. Depending on the individual needs of a paper, may encompass more than one page.

Title pages for all submissions **must** include the following items (as shown in the sample Title Page):

1. **Title** (97 characters [including spaces] or fewer)
2. **Author listing.** Full names for all authors, including degrees, and institutional/professional affiliations. These affiliations should list the institution where the research presented in the article took place; if the affiliation has changed, add a note indicating the additional affiliation. If published, author names and affiliations will appear as seen in the submitted manuscript Word document and the final typeset proofs; all authors must ensure that their information is correct. *Pediatrics* permits a statement of equal contribution for two first authors and two senior authors; on the title page, include asterisks by each name and a statement that reads: * *Contributed equally as co-first authors* or * *Contributed equally as co-senior authors.*
3. **Corresponding Author.** Contact information for the Corresponding Author (including: name, address, telephone, and e-mail). Note that the affiliation should list

the institution where the research presented in the article took place; if the affiliation has changed, add a note indicating the additional affiliation. *Pediatrics* allows one Corresponding Author only; the position of Corresponding Author does not imply seniority or any other status.

4. **Short title** (55 characters [including spaces] or fewer). Please note: The short title may be used on the cover of the print edition.
5. **Conflict of Interest Disclosures** for all authors. This includes any potential conflicts of interest, any relevant financial relationships, and any other relationships or activities that could be perceived to have influenced the work. If none, say "The authors have no conflicts of interest relevant to this article to disclose."
6. **Funding/Support**. Research or project support, including internal funding, should be listed here; if the project was done with no specific support, please note that here. Technical and other assistance should be identified in Acknowledgments. If your funding body has open access requirements, please contact the Editorial Office prior to submission. *Pediatrics* offers Green OA and Gold OA options.
7. **If applicable, Clinical Trial registry name, registration number, and data sharing statement**. We adhere to ICMJE guidelines, which require that all trials must be registered with ClinicalTrials.gov or any other WHO Primary registry. All articles reporting results of clinical trials must also include the Data Sharing Statement.
8. **Abbreviations**. List and define abbreviations used in the text. If none, say "Abbreviations: none".
9. **Article Summary.** All articles with abstracts require this summary. This brief summary is limited to 25 words. For accepted manuscripts, this will appear under the author names in the table of contents to give the reader a brief insight into what the article is about. It should entice the reader to read the full article. For example: *"Through linkage of state Medicaid and Child Protective Services databases, this study captures similarities and differences in health care expenditures based on a history of child maltreatment."*
10. For Regular Article submissions, include both the **"What's Known on This Subject"** and the **"What This Study Adds"** summaries (see below under Regular Article type for description). These are not needed for any other article type.

If a title page does not include all of the above items, the submission may be returned to the authors for completion.

1.6 CONTRIBUTORS' STATEMENT PAGE

All submissions (excluding Commentaries) must contain a Contributors' Statement Page, directly following the Title Page(s) and in the specific format described below. Manuscripts lacking a properly formatted Contributors' Statement Page will be returned to the authors for correction. If you select double-blind peer review, the Contributors' Statement Page should be part of your separate Title Page file.

All persons designated as authors must qualify for authorship (see "Publication Ethics" above), and all those who qualify should be listed. Each author should have participated sufficiently in the work to take public responsibility for appropriate portions of the content. The Contributors' Statement Page lists the authors and specifies the contribution(s) made by each individual. If multiple individuals have identical contributions they may be listed together; do not list an author more than once.

You must follow the required format when creating your Contributors' Statement Page or your manuscript will be returned for correction.

- Each author should only appear once.

- Use names, not initials.
- If multiple authors have identical contributions, you can list them in the same sentence; otherwise, list each author separately.
- Conclude your statement by confirming that: All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

Sample Contributors' Statement:

Dr Smith and Prof Jones conceptualized and designed the study, drafted the initial manuscript, and reviewed and revised the manuscript.
Drs Brown, Grey, and Black and Ms Johnson designed the data collection instruments, collected data, carried out the initial analyses, and reviewed and revised the manuscript.
Dr Green conceptualized and designed the study, coordinated and supervised data collection, and critically reviewed the manuscript for important intellectual content.
All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

Note:

Acquisition of funding, collection of data, or general supervision of a research group alone does not constitute a sufficient basis for authorship. Contributors who do not meet the criteria for authorship (such as persons who helped recruit patients for the study, or professional editors) should be listed in an Acknowledgments section placed after the manuscript's conclusion and before the References section. Because readers may infer their endorsement of the data and conclusions, these persons must give written permission to be acknowledged. These permissions do not need to be submitted with the manuscript unless requested by the editors.

1.7**WORD COUNT**

To determine article length, count the body of the manuscript (from the start of the Introduction to the end of the Conclusion). The title page, contributors' statement page, abstract, acknowledgments, references, figures, tables, and multimedia are not included.

1.8**FIGURES, TABLES, AND SUPPLEMENTARY MATERIAL**

For any figure, table, or supplementary material reproduced or adapted from another source, authors are required to obtain permission from the copyright holder, and proof of permission must be uploaded at the time of submission. The legend must include a statement that the material was used or adapted with permission.

1.8.1Figures

Authors should number figures in the order in which they appear in the text. Figures include graphs, charts, photographs, and illustrations. Each figure must include a legend (placed as a list appearing after the References) that does not exceed 50 words. Abbreviations previously expanded in the text are acceptable. Upload figures as separate files; list figure legends as the last item in your main Word/text file. *Do **not** paste figures into your manuscript text/Word file.* There is no maximum number for figures.
Figure arrays should be clearly labeled, preassembled, and submitted to scale. Figure parts of an array (A, B, C, etc.) should be clearly marked in capital letters in the upper left-hand corner of each figure part.

Technical requirements for figures: The following file types are acceptable: TIFF, PDF, EPS, and PNG. Color files must be in CMYK (cyan, magenta, yellow, black) mode. Pediatrics ***cannot*** accept Excel or PowerPoint files for any part of your submission. To repeat:

upload figures as separate files; list figure legends as the last item in your main Word/text file. *Do **not** paste figures into your manuscript text/Word file.*
Style for figures: Readers should be able to understand figures without referring to the text. Avoid pie charts, 3-dimensional graphs, and excess ink in general. Make sure that the axes on graphs are labeled, including units of measurement, and that the font is large enough to read. Generally delete legends or other material from the graph if it makes the picture smaller. Color graphs should be interpretable if photocopied in black and white.

1.9JOURNAL STYLE

All aspects of the manuscript, including the formatting of tables, illustrations, and references and grammar, punctuation, usage, and scientific writing style, should be prepared according to the most current *AMA Manual of Style* (http://www.amamanualofstyle.com).[1]
Author Listing. All authors' names should be listed in their entirety, and should include institutional/professional affiliations and degrees held. If published, author names and affiliations will appear as seen in the submitted manuscript Word document and the final typeset proofs. All authors must ensure that their information is correct.
Authoring Groups. To include an authoring group, note the following to ensure all individuals are correctly acknowledged:

- On the title page, list any study group at the end of the author listing, preceded either by "for" or "on behalf of" and followed by an asterisk, such as: "on behalf of the XXXXX Study Group*."
- Add the following line: "*A complete list of study group members appears in the Acknowledgments." (Alternatively, list these non-author contributors in an Appendix and change the statement accordingly.)
- Ensure that your Acknowledgments section, placed at the end of the main text and before the References, clearly delineates group members and non-group members. For example: "Members of Study Group ABC include: Person 1, Person 2, Person 3, Person 4. We also wish to acknowledge Person 5 and Person 6 for technical editing."
- **Do not** list the group name as an author in the online metadata section. Only authors who fulfill all four authorship criteria should be included in the online metadata, the title page, and the contributors statement page. These authors should not be included in the acknowledgments, which are reserved for thanking non-author contributors.

Titles. *Pediatrics* generally follows the guidelines of the *AMA Manual of Style* for titles. Titles should be concise and informative, containing the key topics of the work. Declarative sentences are discouraged as they tend to overemphasize a conclusion, as are questions, which are more appropriate for editorials and commentaries. Subtitles, if used, should expand on the title; however, the title should be able to stand on its own. It is appropriate to include the study design ("Randomized Controlled Trial"; "Prospective Cohort Study", etc.) in subtitles. The location of a study should be included only when the results are unique to that location and not generalizable. Abbreviations and acronyms should be avoided. The full title will appear on the article, the inside table of contents, and in MEDLINE. Full titles are limited to 97 characters, including spaces. Short titles must be provided as well and are limited to 55 characters, including spaces. Short titles may appear on the cover of the journal as space permits in any given issue.
Abbreviations. List and define abbreviations on the Title Page. Unusual abbreviations should be avoided. All terms to be abbreviated in the text should also be spelled out at first

mention, followed by the abbreviation in parentheses. The abbreviation may appear in the text thereafter. Abbreviations may be used in the abstract if they occur 3 or more times in the abstract. Abbreviations should be avoided in tables and figures; if used they should be redefined in footnotes.

Units of Measure. Like many US-based journals, *Pediatrics* uses a combination of Système International (SI)[2,3] and conventional units. Please see the *AMA Manual of Style* for details.

Proprietary Products. Authors should use nonproprietary names of drugs or devices unless mention of a trade name is pertinent to the discussion. If a proprietary product is cited, the name and location of the manufacturer must also be included.

Page/Line Numbers. Include page numbers and line numbers in your manuscript so that editors and peer reviewers may more easily point out items/revisions. Line numbers may be continuous or restart on each page.

References. Authors are responsible for the accuracy of references. Citations should be numbered in the order in which they appear in the text. Reference style should follow that of the *AMA Manual of Style*, current edition. Abbreviated journal names should reflect the style of Index Medicus. Visit: https://www.ncbi.nlm.nih.gov/nlmcatalog/journals. If you used reference management software (eg, EndNote) to prepare your manuscript, you must convert the file to plain text prior to submission. Please note: The generated HTML page with the References is for staff/editorial use only; please do not use Word line numbering on your references.

References

1. Iverson C, Christiansen S, Flanagin A, et. al. *AMA Manual of Style*. 10th ed. New York, NY: Oxford University Press; 2007.
2. Lundberg GD. SI unit implementation: the next step. *JAMA*. 1988;260:73-76.
3. Système International conversion factors for frequently used laboratory components. *JAMA*. 1991;266:45-47.

CASE REPORT

Abstract length: 250 words or fewer (unstructured: no headings, run in a single paragraph)

Article length: 1,600 words or fewer

Author limit: Seven (7) authors or fewer (with rare exception)

Case Reports highlight unique presentations or aspects of disease processes that may expand the differential diagnosis and improve patient care. In general, case reports will include 10 cases or fewer. For a manuscript to be considered a Case Report, it must meet at least one of the following three criteria:

1. Challenge an existing clinical or pathophysiologic paradigm.
2. Provide a starting point for novel hypothesis-testing pre-clinical or clinical research.
3. Focus on topics pertinent to the pediatric generalist, allowing pediatrics colleagues to provide improved care. (Manuscripts meeting this criterion will be prioritized over other submissions.)

Case Reports should consist of an unstructured abstract that summarizes the case(s), a brief introduction (recommended length, 1-2 paragraphs), a section that details patient presentation, initial diagnosis and outcome, as well as a discussion that includes a brief review of the relevant literature and describes how this case brings new understanding about the presentation, diagnostic approach, and/or novel treatment of a disease. Case Reports that merely present, for example, the third published case of a clinical condition, that describe a

patient who has 2 rare conditions, or that detail the youngest patient with a well-described disease do not on those merits alone meet the bar for publication in *Pediatrics.*
Authors may find the criteria for case reports as contained in the CARE guidelines useful in preparing their manuscript.
Written consent must be obtained from the parent or guardian. You do not need to include a copy with your submission unless the patient may be identifiable; however, a copy must be provided to *Pediatrics* upon request. *Pediatrics* does not supply a consent form.
The general submission instructions (including cover letter, title page, contributors' statement page, journal style guidance, and conflict of interest statements) also apply to Case Reports.
Do **not** include "a case report" or similiar language in your title as this is redundant; published manuscripts will appear in the Case Reports section.

PARECER DO COMITÊ DE ÉTICA

UNIVERSIDADE FEDERAL DE SANTA CATARINA - UFSC

PARECER CONSUBSTANCIADO DO CEP

DADOS DO PROJETO DE PESQUISA

Título da Pesquisa: Mucinose folicular na infância: um diagnóstico raro
Pesquisador: Maria Marlene de Souza Pires
Área Temática:
Versão: 1
CAAE: 48289021.9.0000.0121
Instituição Proponente: CENTRO DE CIÊNCIAS DA SAÚDE
Patrocinador Principal: Financiamento Próprio

DADOS DO PARECER

Número do Parecer: 4.822.925

Considerações sobre os Termos de apresentação obrigatória:
Folha de rosto assinada pelo pesquisador responsável e pelo coordenador do Curso de Graduação em Medicina, professor Aroldo Prohmann de Carvalho.

A apresentação do caso, na forma de artigo, contribuiu para que o CEP/UFSC tenha concordado com o pedido de dispensa do TCLE, visto que não é possível qualquer identificação do participante.

Recomendações:
Não há.

Conclusões ou Pendências e Lista de Inadequações:
Aprovado.

Situação do Parecer:
Aprovado

Necessita Apreciação da CONEP:
Não

FLORIANOPOLIS, 02 de Julho de 2021

Assinado por:
Luciana C Antunes
(Coordenador(a))