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REVIEW PROTOCOL

Shoulder musculoskeletal changes present in Sprengel deformity: a scoping review protocol

Alterações musculoesqueléticas do ombro presentes na deformidade de Sprengel: um protocolo de revisão de escopo

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Abstract

Introduction: Sprengel syndrome is a rare condition in which the scapula is located in an abnormally high position on the back. This condition occurs due to abnormal musculoskeletal development in the shoulder region during pregnancy. Objective: to verify in the literature which musculoskeletal changes in the shoulder complex are present in children with Sprengel syndrome. Methods: a scoping review of primary, observational or interventional studies will be carried out, as long as they report musculoskeletal changes in patients with Sprengel deformity. The study protocol was registered on the Prospero Platform (CRD42022381782). We will include children aged 5 to 10 years with Sprengel's deformity (even if a longer period). Exclusion criteria: any other type of congenital comorbidity not associated with Sprengel's deformity. The searches will be carried out in the following databases: Medical Literature Analysis and

Retrieval System Online (Medline) via Pubmed, Excerpta Medica dataBASE (Embase) via Elsevier, Literatura Latino-Americana, Scientific Electronic Library Online (SciELO) and the Caribbean in Health Sciences (Lilacs) via the Virtual Health Library Portal, using relevant descriptors and synonyms. Two authors will extract the data independently and possible disagreements will be resolved with a third author. Evidence will be presented according to the Joanna Briggs Institute (JBI) method. Expected results: Map the musculoskeletal changes in the shoulder complex present in Sprengel's deformity to assist professionals in clinical practice and support future studies on this condition.
Keywords: Sprengel deformity; congenital diseases; musculoskeletal syndromes.

Resumo

Introdução: A síndrome de Sprengel é uma condição rara em que a escápula está localizada em uma posição anormalmente alta no dorso. Essa condição ocorre devido a um desenvolvimento anormal musculoesqueléticas na região do ombro durante a gestação. Objetivo: verificar na literatura quais alterações musculoesqueléticas do complexo do ombro estão presentes em crianças com a síndrome de Sprengel. Métodos: será realizada uma revisão de escopo de estudos primários, observacionais ou intervencionais, desde que relatam alterações musculoesqueléticas em pacientes com deformidade de Sprengel. O protocolo do estudo foi registrado na Plataforma Próspero (CRD42022381782). Incluiremos indivíduos crianças de 5 a 10 anos com deformidade de Sprengel (mesmo que um período mais longo). Critérios de exclusão: qualquer outro tipo de comorbidade congênita não associada a deformidade de Sprengel. As buscas serão realizadas nas bases de dados: *Medical Literature Analysis and Retrieval System Online (Medline)* via Pubmed, *Excerpta Medica dataBASE (Embase)* via Elsevier, Literatura Latino-Americana, *Scientific Electronic Library Online (SciELO)* e do Caribe em Ciências da Saúde (Lilacs) via Portal da Biblioteca Virtual em Saúde, utilizando descritores e sinônimos relevantes. Dois autores irão extrair os dados de forma independente e possíveis divergências serão resolvidas com um terceiro autor. As evidências serão apresentadas de acordo com o método de *Joanna Briggs Institute (JBI).* Resultados esperados: Mapear as alterações musculoesqueléticas do complexo do ombro presentes na deformidade de Sprengel para auxiliar profissionais da prática clínica e embasar estudos futuros sobre esta condição.
Palavras-chave: deformidade de Sprengel; doenças congênitas; síndromes musculoesqueléticas.

Introduction

The shoulders are essential for human functionality. Their specific biomechanics make them the most mobile joint complex in the body, providing the upper limbs with mobility in three axes of space [1]. Frequent shoulder dysfunctions are the third leading cause of musculoskeletal consultations in primary healthcare [2].

Congenital high scapula, also known as Sprengel's deformity or undescended congenital scapula, is uncommon but ranks among the most frequent congenital abnormalities of the shoulder [3]. Embryologically, this congenital deformity arises from an interruption in the normal caudal migration of the scapula between the ninth and 12th weeks of

intrauterine life, resulting in a raised, mal-rotated, and hypoplastic position of the scapula, surrounded by hypoplastic periscapular muscles, which hold the scapula and further limit rotation [4].

In Sprengel's deformity, the scapula can be elevated between 2 and 10 cm and adducted, with its lower angle rotated medially, often approaching the midline. Due to this medial rotation, the glenoid cavity faces downward, and the superomedial angle of the scapula is rotated upwards, forming a characteristic prominence in the suprascapular region, causing fullness on the ipsilateral side of the neck, with a loss of the normal contour of the shoulder [5].

The main clinical alterations involve the abnormal positioning and hypoplasia of the scapula, which can result not only in aesthetic issues but also in limitations of movements in the shoulder girdle [6]. Furthermore, several authors point out that there may be the presence of the omovertebral bone, which connects the scapula to the vertebral column [3,7–9].

In terms of the surgical treatment of Sprengel's deformity, the primary objective would be to improve aesthetics, which in itself does not guarantee functional improvements. Therefore, several surgical procedures have been developed to enhance functional outcomes [10]. Among the surgical procedures for correcting Sprengel's deformity found in the literature, the Woodward procedure and the modified Green procedure stand out.

However, despite the literature describing orthopedic surgical treatment methods for this condition, there is still a scarcity of scientific evidence regarding the functional rehabilitation and musculoskeletal alterations in patients with Sprengel's deformity, as well as a lack of information on the quality of life of this population over time.

In light of the above, the objective is to map the scientific evidence regarding musculoskeletal alterations in children aged 5 to 10 years with Sprengel's syndrome. This aims to assist researchers and clinicians in understanding biomechanical alterations, assessment tools, and to highlight knowledge gaps to support future studies.

Identifying the research question

The aim of this review is to map musculoskeletal alterations in children aged 5 to 10 years with Sprengel's syndrome. Therefore, the following research questions are proposed to be addressed:

1. What are the musculoskeletal alterations present in Sprengel's syndrome?
2. What instruments are used to assess functional capacity and quality of life in patients with Sprengel's syndrome?
3. "What interventions are practiced in Sprengel's syndrome?"

Methods

Type of study

This study is a protocol for a scoping review that was developed following the recommendations proposed by the Preferred Reporting Items for Systematic Reviews and Meta-Analyses extension for Scoping Reviews (PRISMA-ScR).

Ethical aspects and research location

The review protocol has been registered on the Prospero Platform (CRD42022381782). This study will be conducted at the Federal University of Amapá (UNIFAP), in the undergraduate program of Physiotherapy, Department of Biological and Health Sciences.

Eligibility criteria

Types of included studies

We will include primary studies, observational or interventional, published either in full-text or as abstracts.

Types of participants

Individuals aged 5 to 10 years with Sprengel's deformity (even if long-term follow-up has been conducted). We will exclude any congenital comorbidities unrelated to Sprengel's deformity.

Types of interventions

Studies assessing musculoskeletal alterations, functional capacity, quality of life in the shoulder complex in children with Sprengel's deformity. Additionally, studies reporting the type of intervention applied, whether physiotherapeutic or surgical.

Collection procedure

Literature search strategy

We will conduct sensitive searches using pre-established relevant terms and descriptors, without limitations on publication year or language, in the following databases:

- Medical Literature Analysis and Retrieval System Online (MEDLINE) via PubMed;
- Excerpta Medica dataBASE (Embase) via Elsevier;
- Latin American and Caribbean Health Sciences Literature (LILACS) via the Virtual Health Library Portal;
- Scientific Electronic Library Online (SciELO);

Additionally, we will analyze the reference lists of relevant publications. Finally, we will reach out to the authors of primary studies to identify potentially important additional studies for this review and request further information if necessary."

Database	Search strategy
MEDLINE through PubMed	((“Sprengel deformity” OR undescended scapula OR High scapula OR Sprengel’s OR Waaler Aarskog OR “Poland Syndrome” OR Klippel-Feil OR “ribs Abnormalities” OR omovertebral bone OR “Congenital high scapula”) AND (infan* OR pre school OR pre-school OR child*))
Embase through Elsevier	#1 (‘sprengel deformity’/exp OR ‘undescended scapula’ OR ‘high scapula’ OR sprengel`s OR ‘waaler aarskog’ OR ‘poland syndrome’/exp OR ‘poland syndrome’ OR ‘poland`s syndrome’ OR ‘polands syndrome’ OR ‘poland syndactyly’ OR ‘poland`s syndactyly’ OR ‘klippel feil syndrome’/exp OR ‘ribs abnormalities’/exp OR ‘omovertebral bone’ OR ‘congenital high scapula’) AND (‘infant’/exp OR ‘infant’ OR ‘pre school’ OR ‘child’/exp OR ‘child’ OR ‘children’) #2 #1 AND [embase]/lim NOT ([embase]/lim AND [medline]/lim)
Scientific Electronic Library Online (SciELO)	((‘sprengel deformity’))
Latin American and Caribbean Health Sciences Literature (LILACS) via the Virtual Health Library Portal	((sprengel deformidade) OR (escápula alta) OR sprengel’s OR waaler aarskog OR (síndrome de poland) OR klippel-feil OR (anormalidade das costelas) OR (osso omovertebral)) AND (infan* OR pre escola* OR criança* OR mh:criança OR mh:m01.060.406*) AND (db:(“LILACS” OR “IBECS” OR “BINACIS” OR “CUMED” OR “LIPECS” OR “BBO” OR “BRISA”))

Table 1 - *Scoping Review Search Strategy*

Study selection

Two authors (E.A.P.) and (P.G.L.) will independently conduct the study selection based on predefined eligibility criteria. Initially, studies indexed in more than one database (duplicates) will be excluded. Then, evaluation will be based on titles and abstracts, followed by a thorough examination of the full texts for a more precise analysis. Any disagreements between the authors regarding study inclusion will be resolved by a third author (I.L.). To enhance the selection process, we will use the Rayyan application (https://www.rayyan.ai/). The mapped results will be presented narratively accompanied by a flowchart, following the PRISMA recommendation for the Scoping Review process.

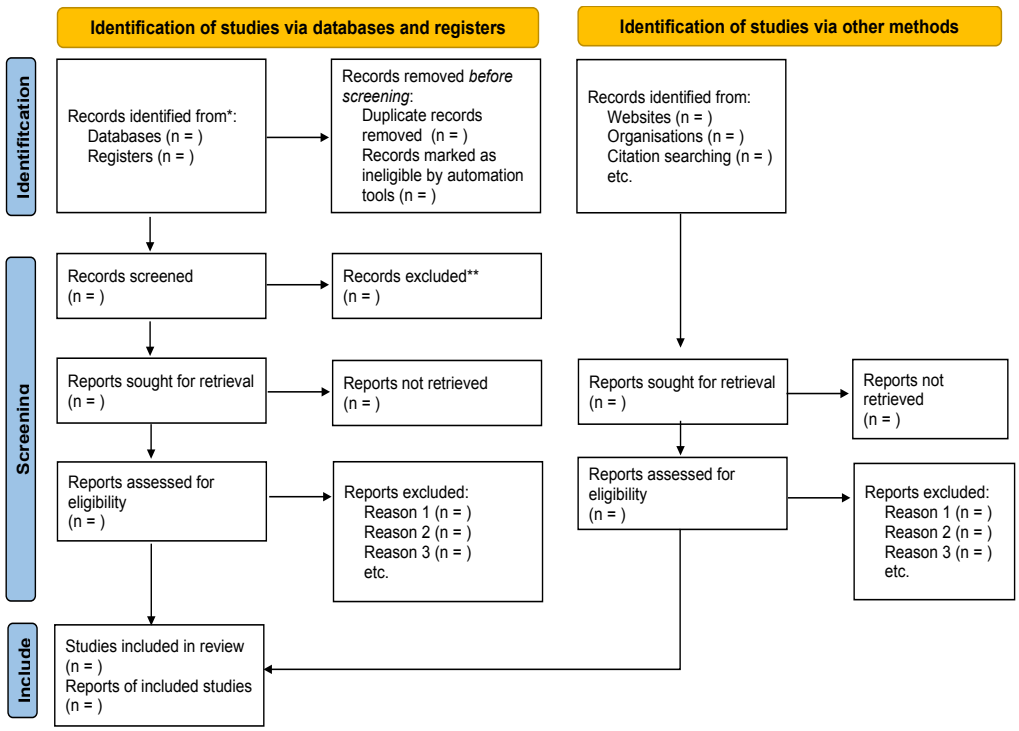

Figure 1: *Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) Flow Diagram for the Scoping Review Process.*

Extracting and Mapping the Results

Two authors (E.A.P.) and (P.G.L.) will independently extract the found data. The results will be presented in tables addressing the research questions with the following variables: 1) Study identification details (titles, authors, study location); 2) Participants: number of participants in the study, age, age range, gender, diagnosis duration, inclusion criteria, and exclusion criteria; 3) Musculoskeletal alterations; 4) Instruments used for functional capacity assessment; 5) Instruments used for quality of life assessment; 6) Interventions: type of intervention (surgery or physiotherapy). These results will be presented following the Joanna Briggs Institute (JBI) methodological recommendations, synthesized as a 'map' to best convey the findings of this scoping review.

Impacts and expected results

The present study aims to conduct a scoping review to map the musculoskeletal alterations in the shoulder complex present in Sprengel's deformity. This review seeks to identify and describe the anatomical changes occurring in the shoulder complex in individuals with Sprengel's deformity. This may include anomalies in the position and shape of the scapula, displacement of the clavicle, alterations in adjacent muscles and ligaments, among other aspects. This detailed characterization of anatomical changes will provide a solid foundation for future research, enabling a better understanding of the pathophysiology of the deformity.

Furthermore, it is expected to map the instruments used to assess functional capacity and quality of life in these patients, offering an overview of these tools for researchers and professionals in clinical practice. Identifying gaps in the current scientific literature regarding shoulder complex alterations in individuals with Sprengel's deformity is crucial. By mapping available evidence through this scoping review, researchers can direct efforts to fill these gaps with more in-depth and high-quality studies. This may involve the need for longitudinal research, randomized controlled clinical trials, and studies evaluating different therapeutic approaches, guiding future high-quality research.

Conflicts of interest

We declare no conflicts of interest.

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Authors contribution

Conception and design of the research: Iosimuta NCR, Pinto ACPN, Alves, INL; Data collection: Teixeira EAP, Demes PGO, Lopes I, Távora ASA; Data analysis and interpretation: Teixeira EAP, Demes PGO, Lopes I; Statistical analysis: Iosimuta NCR, Pinto ACPN; Manuscript writing: Teixeira EAP, Demes PGO, Lopes I; Critical revision of the manuscript for important intellectual content: Teixeira EAP, Demes PGO, Lopes I, Távora ASA, Iosimuta NCR, Pinto ACPN.

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