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Title: The care and management of children and young people with ataxia telangiectasia provided by nurses and allied health professionals: a scoping review protocol

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Introduction

Ataxia Telangiectasia (AT) is a rare, genetic and progressive condition, usually presenting in early childhood and primarily affecting the neurological, immunological and pulmonary systems (Rothblum-Oviatt *et al.*, 2016). Children with AT typically present with cerebellar ataxia in early childhood, generally before the age of four years, with oculomotor, extrapyramidal and peripheral nervous system problems occurring in later childhood and adolescence (van Os *et al.*, 2017). Difficulties with feeding, swallowing and nutrition are common (Ross *et al.*, 2015). World-wide prevalence estimates vary between 1 in 40,000 and 1 in 100,000 live births (Rothblum-Oviatt *et al.*, 2016). People with AT are at high risk of respiratory tract infections and malignancy (Reiman *et al.*, 2011). Cause of death in the second or third decade of life is usually due to respiratory failure or cancer (Crawford *et al.*, 2006, van Os *et al.*, 2017). In the absence of a cure, people living with AT require co-ordinated multi-disciplinary care to optimally manage their complex needs (van Os *et al.*, 2017). Symptomatic management and rehabilitation can greatly improve quality of life and prevent complications that could increase morbidity and mortality (Perlman *et al.*, 2011).

The available guidance on the management of children and young people living with AT has largely focused on diagnosis and medical interventions with only limited provision for the nursing and allied health professions (for example Taylor *et al.*, 2014; van Os *et al.*, 2017). The views of children and their parents/carers have been given little attention even though this complex condition raises significant medical, physical and psychosocial issues for children and families (Capra, 2006). Whilst specialist centres provide expert clinical support for people with rare disorders, such as AT, allied healthcare is typically provided by local community healthcare teams, where knowledge of rare conditions is generally low (von der Lippe *et al.*, 2017). Access to high quality evidence-led care across services is essential to ensure that people with rare conditions are not disadvantaged (Department of Health, 2018). Comprehensive guidance for nurses and allied health care teams,

based on a systematic search of multiple sources of evidence, is therefore needed to improve knowledge and to share expertise across specialist, acute, community and rehabilitation settings.

To address this need, we plan to undertake a scoping review of the care and management of children and young people with AT provided by nurses and allied health professionals (AHPs). Unlike systematic reviews that tend to address relatively precise questions, scoping reviews can be used to map and summarize the evidence from a diverse field of literature and provide an overview of what evidence has been produced regardless of quality (Peters *et al.*, 2020). We aim to map and establish the scope of relevant literature, identify and analyse knowledge gaps to inform practice and further research.

This scoping review will seek to answer the following question:

- What guidance is available to inform the care and management of children and young people with ataxia telangiectasia from the perspective of nursing and allied health professions?

This scoping review aims to:

- Systematically identify and map the type, range, scope and content of published and unpublished studies, reviews, guidelines, guidance documents and practice documents concerning nursing and allied health care and management of children and young people with AT.

This is the first review that seeks to identify and map the literature concerning the nursing and allied health care management of children and young people with AT. Preliminary investigations have indicated that evidence guiding the care and management of AT is limited, which is likely to be because AT is currently under-researched, like many rare and complex conditions. Therefore, we will also search for evidence from other childhood conditions, known to share some significant pathophysiology, such as cerebral palsy and muscular dystrophy with the intention, where feasible, to translate relevant knowledge to the AT population. This scoping review has been registered on the Centre for Open Science where the registration form is publicly accessible.

Methods

The Joanna Briggs Institute Manual for Scoping Reviews (Peters *et al.*, 2020) and the Preferred Reporting Items for Systematic Reviews and Meta-analyses extension for Scoping Reviews (PRISMA-ScR) (Tricco *et al.*, 2018) have been used to inform the review methods. Preliminary searches of conventional healthcare databases indicated that sources of evidence may be hard to find for AT

specific nursing and allied health interventions. As we therefore lack a focused line of enquiry, a broad search strategy will be adopted using a wide range of sources to explore the topic.

Inclusion Criteria

Population

This scoping review will consider studies, reviews, guidelines, guidance documents and practice documents whose patient population consists of children and young people up to the age of 24 years with nervous system diseases. The upper age limit is in accordance with the United Nations definition of a child and youth (United Nations, 1981). Adults are excluded from this review because AT is primarily a condition affecting children and young people, and therapies or interventions devised for adults are unlikely to generalise to the focus population.

Under nervous system diseases, we would primarily be interested in ataxia or AT. However, our preliminary investigations have shown that there may be very little literature specifically focussed on ataxia or AT. Therefore, other related childhood conditions such as cerebral palsy or muscular dystrophy, will also be included in the search strategy to inform the scoping review. This decision is based on the potential generalisability of knowledge between these conditions and AT. Where sources of evidence include mixed population groups, the decision whether or not to include the study in the review will be case based, according to relevance and the number of children with a relevant condition included in each study.

Concept

We will consider sources of evidence with a focus on care and management provided by nurses and allied health professionals (AHPs). The term 'Allied Health Professionals' is used within the UK to describe a diverse range of 14 autonomous professions (Harris *et al.*, 2019). For the purpose of this study we have included the following AHPs; physiotherapists (neurological and respiratory physiotherapy specialists), occupational therapists, speech and language therapists, dietitians, orthoptists, optometrists, orthotists and podiatrists. These professions were chosen as the focus because they represent the largest groups of therapists and are most likely to be involved in the care of children and young people with AT. For inclusion, care and management practices must focus on prevention, management or treatment of primary impairments, activity limitations, participation restrictions, or secondary health conditions, or interventions for health promotion, or support and enabling independence. Surgical interventions will only be included if nursing and/or any of the selected AHPs are significantly involved in pre or post-surgical care. Medical and pharmacological interventions and practices will be excluded. Psychotherapy and talking therapies, such as

counselling, are also excluded as these present a significant and complex class of intervention, which is currently outside of the scope of this project.

We will identify and map the following outcomes:

Primary outcomes: Activity, participation and health-related quality of life

Secondary outcomes: Impairment level measures related to neuromusculoskeletal performance, cardiovascular performance, lung function, nutritional status, swallow, and oculo-motor function.

For qualitative studies we will report subjective perspectives about the healthcare experiences of children and young people with AT (or related conditions) or their parents/carers and subjective experiences of nurses and AHPs working with children and young people with AT or related conditions.

Context

There will be no restrictions on the healthcare setting, geographical location or cultural group. No specific constraints will be placed on where care and management is delivered, or whether it is delivered via a single discipline or by multi-disciplinary care teams (that include at least one nurse or AHP), or whether it is delivered to individuals or groups of children and young people with AT or related conditions.

Types of evidence sources

For the purpose of this review, all types of evidence including published and unpublished research studies of any type (e.g. randomised controlled trials, before and after studies, case studies and case series, qualitative studies), partially published research studies of any type (e.g. abstracts), reviews, guidelines, guidance documents, practice documents, and grey literature sources will be included. For quantitative intervention studies where comparisons are made, we will include comparisons of no treatment or usual care or one treatment versus another. No date or language limits will be applied to the search.

Search strategy

A comprehensive search strategy has been developed in consultation with an information retrieval specialist and a team of experts in their respective fields. Consistent with guidelines recommended in all JBI types of reviews (Peters *et al.*, 2020), a three-step search strategy will be utilized. Firstly, an initial limited search of two databases, CINAHL and PubMed, will be conducted with the assistance of an information retrieval specialist. At this stage, the title and abstracts of the retrieved sources of evidence will be reviewed. The full version of the proposed search strategy for PubMed is detailed in

Appendix 1. Where we anticipate limited AT or ataxia specific evidence sources, we have used the MeSH term 'Nervous System Diseases', to capture evidence sources for conditions such as cerebral palsy or muscular dystrophy. For professions such as physiotherapy where we believe there will be more sources of evidence we have used the MeSH term 'Ataxia'. This decision will be revised if the results indicate that broader search will need to be undertaken.

The second step will include extension of the search query to the other relevant electronic databases such as Essential Evidence Plus, Cochrane Central Register of Controlled Trials, Joanna Briggs Institute (JBI) EBP Database and PEDro. A complete list of the sources of information including the grey literature sources is detailed in Appendix 2. At this stage too, the title and abstracts will be reviewed for selection. Thirdly, the reference list of the selected sources of evidence will be screened for inclusion. Where possible, citation tracking will be used for all included sources of evidence. Where necessary, authors of included publications will be contacted for further information.

Study selection

Study selection will be carried out by two independent reviewers. Based on the inclusion criteria, the reviewers will review the title and abstracts of the retrieved search results. To ensure consistency in study selection between the two reviewers, a sample of 25 titles and abstracts will be selected. The reviewers will screen these using the eligibility criteria and the results will then be compared. If required, any discrepancies will be discussed and modifications in the inclusion criteria will be made. Study selection will then be carried out independently by the two reviewers using the modified inclusion criteria. Firstly, all search results will be screened for potential inclusion based on the examination of titles and abstracts. Secondly, full text of all selected sources of evidence will be screened for inclusion in the review. Disagreements will be solved by consensus or by the decision of a third reviewer (Peters *et al.*, 2020).

To expedite the screening and selection process of the sources of evidence, a web-tool called Rayyan QCRI (Ouzzani *et al.*, 2016) will be used at all stages of the study selection process.

The search for this scoping review will be iterative as reviewers will become more familiar with the evidence base, additional keywords and sources, and potentially useful search terms may be discovered and incorporated into the search strategy (Peters *et al.*, 2020). Any deviation from the protocol will be identified and reported in the main scoping review report. The search will be updated prior to publication to identify new sources of evidence.

Data charting

Data from the selected literature will be charted by the reviewers using a data charting table adapted from the JBI reviewer's manual (Peters *et al.*, 2020; see appendix 3). This table has been pre-pilot tested on two publications (one guidance document and one research study) by both the reviewers to ensure it records all the key information of the sources of evidence. The data charting process will be undertaken by both the reviewers where one reviewer will chart and the other reviewer will verify the data. To ensure all relevant results are charted, the charting table will again be pilot tested on the first five publications and amended as necessary. This pilot step will also be conducted by both reviewers. The final version of the data charting table will be included in the full scoping review report.

Analysis of Evidence

The occurrence of concepts, characteristics, populations and other data fields with simple frequency counts will be investigated (Peters *et al.*, 2020). The qualitative data extracted from the sources of evidence will be mapped descriptively by undertaking content analysis. Figures and charts may be used to support the analysis and to represent the data.

Presentation of the results

The PRISMA flowchart (Moher *et al.*, 2009) will be used to report the search process and the final number of sources of evidence included along with the reasons for exclusion in the full scoping review report. The included sources of evidence will be presented in a table (see appendix 4). The tabular presentation of the results will be accompanied by a narrative summary to describe how the results relate to the review aims and question (Peters *et al.*, 2020). It is anticipated that the evidence will report upon a diverse range of care and management services provided by nurses and AHPs, and as such the sources of evidence will be grouped under five broad themes; swallow, speech, respiratory, exercise and function, and nutrition. These themes reflect the multidisciplinary approach used by AHPs and nurses to address the common problems faced by people with AT and related conditions. The primary focus of the review is to present findings related to the care and management of AT. However, comment will also address results related to other types of ataxia and other related nervous system diseases, where relevant. Results for each theme will be discussed and presented clearly in the full scoping review report.

Consultation with relevant stakeholders

The consultation stage provides opportunities for stakeholder involvement, providing insights beyond what is reported in the literature (Levac *et al.*, 2010). The following stakeholders will be engaged:

- We plan to consult with children and young people with AT and their parents/guardians: To discuss the results from the review and facilitate generation of implications for future practice.
- Nurses, AHPs and other relevant healthcare professionals in acute and community sectors: Consulted in developing the search strategy, interpretation of results and analysis and dissemination of results. To provide insight beyond what was reported in the sources of evidence.
- AT specialists and allied professionals at an AT specialist clinic based at Nottingham City Hospital in the UK: To provide insight from an acute sector specialist service.

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Appendix 1: Search strategy for PubMed

Allied health profession or nursing group category	Search terms
Physiotherapy	(Ataxia[mh] OR ataxia*[tw]) AND (Physical Therapy Modalities[mh] OR Circuit-Based Exercise[mh] OR Postural Balance[mh] OR Virtual Reality[mh] OR Virtual Reality Exposure Therapy[mh] OR physiotherap*[tw] OR physical therap*[tw] OR exercise[tw] OR physical activit*[tw] OR training[tw] OR

	coordination training*[tw] OR co-ordination training*[tw] OR coordinative training*[tw] OR co-ordinative training*[tw] OR balance training*[tw] OR proprioceptive training*[tw] OR core stability training*[tw] OR gait training*[tw] OR postural control*[tw] OR postural balance*[tw] OR virtual reality*[tw] OR walking aid*[tw])
Respiratory physiotherapy	(Nervous System Diseases[mh] OR nervous system disease*[tw] OR ataxia*[tw]) AND (Breathing Exercises[mh] OR Drainage, Postural[mh] OR respiratory muscle training*[tw] OR respiratory physiotherap*[tw] OR breathing exercise*[tw] OR breathing pattern*[tw] OR breathing technique*[tw] OR postural drainage*[tw] OR pulmonary muscle training*[tw] OR ventilatory muscle training*[tw] OR inspiratory muscle training*[tw] OR expiratory muscle training*[tw] OR airway clearance technique*[tw] OR forced expiratory technique*[tw])
Dietetics	(Ataxia[mh] OR ataxia*[tw] OR neurological disabilit*[tw] OR neurological impairment*[tw] OR neurologic disabilit*[tw] OR neurologic impairment*[tw] OR neurologically disab*[tw] OR neurologically impair*[tw]) AND (Diet Therapy[mh] OR Nutrition Therapy[mh] OR Nutritional Status[mh] OR Diet, Food, and Nutrition[mh] OR Enteral Nutrition[mh] OR Gastrostomy[mh] OR Energy Intake[mh] OR diet therap*[tw] OR nutrition status*[tw] OR nutritional status*[tw] OR nutrition manag*[tw] OR nutritional manag*[tw] OR nutrition therap*[tw] OR nutritional therap*[tw] OR nutrition support*[tw] OR nutritional support*[tw] OR gastrostomy[tw] OR puree food*[tw] OR non oral feeding*[tw] OR non-oral feeding*[tw] OR enteral nutrition*[tw] OR percutaneous gastrostomy*[tw] OR percutaneous endoscopic gastrostomy*[tw] OR caloric intake*[tw] OR calorie intake*[tw] OR energy intake*[tw])
Speech and language therapy	(Ataxia[mh] OR ataxia*[tw]) AND (Speech Therapy[mh] OR Language Therapy[mh] OR Rehabilitation of Speech and Language Disorders[mh] OR speech*[tw] OR speech therap*[tw] OR language therap*[tw] OR swallow*[tw])
Nursing	(Ataxia[mh] OR ataxia*[tw]) AND (Nursing[mh] OR nurs*[tw])
Occupational therapy	(Ataxia[mh] OR ataxia*[tw]) AND (Occupational Therapy[mh] OR Activities of Daily Living[mh] OR occupational therap*[tw] OR activity of daily living*[tw] OR activities of daily living*[tw])
Orthoptics/Optomety	(Ocular Motility Disorders[mh] OR Ataxia[mh] OR ocular motility disorder*[tw] OR ataxia*[tw]) AND (convergence training*[tw] OR pursuit training*[tw] OR saccade training*[tw] OR visual rehabilitation training*[tw] OR visual scanning training*[tw] OR eye movement training*[tw] OR audiovisual exploration training*[tw])

Orthotics/Podiatry	(Ataxia[mh] OR ataxia*[tw]) AND (Splints[mh] OR Podiatry[mh] OR Orthotic Devices[mh] OR splint*[tw] OR podiatr*[tw] OR orthotic device*[tw] OR orthosis[tw] OR orthoses[tw] OR insole*[tw] OR weighted therap*[tw])
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Appendix 2: Sources of information

a. Electronic databases

1. CINAHL (EBSCOhost)
2. PubMed
3. Cochrane Central Register of Controlled Trials (CENTRAL), part of The Cochrane Library
4. Joanna Briggs Institute (JBI) EBP Database (OVID)
5. PROSPERO
6. Essential Evidence Plus (www.essentialevidenceplus.com/) (includes Cochrane Database of Systematic Reviews)
7. PEDro (www.pedro.org.au/)
8. Health Services Research Projects in Progress (HSRPRoj) (wwwcf.nlm.nih.gov/hsr_project/home_proj.cfm)
9. National Rehabilitation Information Center (www.naric.com/) (NIDILRR Program Database and Knowledgebase only)
10. Clinical trials.gov
11. World Health Organization International Clinical Trials Registry Platform (ICTRP) (www.who.int/ictrp/en/)

b. Grey Literature database

12. Open Grey (<http://www.opengrey.eu/>)

c. Expert opinions

13. Consultation with AT specialists and allied professionals at the children’s clinic in Nottingham, UK
14. Consultation with the Dutch physical activity guidelines authors for children with chronic conditions e.g. chronic respiratory conditions
15. Consultation with any other national and international expert group(s) identified during the literature search

Appendix 3: Data charting table

A. Research study

Scoping Review Details	
1. Scoping Review title:	
2. Review objective/s:	
3. Review question/s:	

Inclusion/Exclusion Criteria	
4. Population	
5. Concept	
6. Context	
7. Types of evidence source	
Evidence source Details and Characteristics	
8. Citation details (e.g. author/s, date, title, type of publication, journal, volume, issue, pages)	
9. Type of evidence source [e.g. randomized control trial, before and after study, qualitative study (include methodology eg. phenomenology, ethnography)]	
10. Aim/Purpose of the evidence source	
11. Country	
12. Language	
13. Context (e.g. setting for intervention delivery, cultural, societal, economic, gender factors)	
14. Participants/population (e.g. age/sex, number, condition, functional ability, other relevant factors to describe the sample)	
15. Diagnostic criteria applied (e.g., ESID/PAGID)	
16. Search source (database/grey literature)	
Details/Results extracted from source of evidence (in relation to the concept of the scoping review)	
17. Name of intervention (quantitative studies)/ Phenomenon of interest (qualitative studies)	
18. Intervention related to profession (e.g. physiotherapy, nursing, etc.)	
19. Inclusion criteria	

20. Exclusion criteria	
For quantitative studies, skip rows 27-28; For qualitative studies, skip rows 21-26	
Quantitative studies only	
21. Mode of delivery (individual, group, supervised, unsupervised)	
22. Target of intervention (e.g. child with ataxia)	
23. Duration of intervention (including frequency and intensity)	
24. Delivery of the intervention	
25. Method of outcome measurement (list all relevant primary and secondary outcomes)	
26. Main outcomes/results reported	
Qualitative studies only	
27. Methods of data collection [e.g., interviews (paired/individual/ other), diaries, focus groups] (also include duration of interviews)	
28. Data analysis (e.g., thematic analysis, framework analysis, IPA)	
All types of studies	
29. Key findings/results	
30. Limitations/Suggestions for further research	
31. Conclusion/Recommendations for practice	
32. Any other comments	

B. Non-research literature (e.g., clinical guidance documents)

Scoping Review Details	
1. Scoping Review title:	
2. Review objective/s:	

3. Review question/s:	
Inclusion/Exclusion Criteria	
4. Population	
5. Concept	
6. Context	
7. Types of evidence source	
Evidence source Details and Characteristics	
8. Citation details (e.g., author/s, date, title, type of publication)	
9. Type of evidence source (e.g., clinical guidance, practice guidance document, grey literature)	
10. Aim/Purpose of the evidence source	
11. Country	
12. Language	
13. Context (e.g., setting, cultural, societal, economic, gender factors)	
14. Population (e.g., age/sex)	
15. Search source (database/grey literature)	
Details/Results extracted from source of evidence (in relation to the concept of the scoping review)	
16. Recommendations for swallow, speech, respiratory, exercise and function, and nutrition	
17. Details of the recommendation	
18. Limitations/Suggestions for further research	
19. Conclusion/Recommendations for practice	
20. Any other comments	

Appendix 4: Data presentation table

Citation details (author/year/country)	Types of evidence source (e.g., research study/review/guidance document)	Population(s) identified (children, young people, parents/caregivers)	Condition identified (e.g., AT, ataxia, cerebral palsy, muscular dystrophy)	Allied health/ nursing profession (e.g., physiotherapy, occupational therapy, dietetics, speech and language therapy)	Themes (swallow, speech, respiratory, exercise and function, and nutrition)