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The Care and Management of Children and Young People with Ataxia Telangiectasia Provided by Nurses and Allied Health Professionals: a Scoping Review

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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**

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~~Supplement 1: A detailed overview of search terms incorporated in the review~~

~~Supplement 2: A list of all the databases that were searched~~

Abstract ~~(250/250 words):~~

Ataxia telangiectasia (A-T) is a rare, multisystem progressive condition that typically presents in early childhood. In the absence of cure, people with A-T require coordinated multidisciplinary care to manage their complex array of needs and to minimize the disease burden. Although symptom management has proven benefits for this population, including improved quality of life and reduced complications, there is a need for guidance specific to the nursing and allied healthcare teams who provide care within the community. A scoping review, adopting the Joanna Briggs Institute methodology, was undertaken. It aimed to identify and map the available expertise from nursing and allied health-care and management of children and young people with A-T ≤18 years of age. A rigorous search strategy was employed which generated a total of 21,118 sources of evidence, of which 50 were selected for review following screening by experts. A range of interventions were identified that reported a positive impact on A-T-related impairments, together with quality of life, indicating that outcomes can be improved for this population. Most notable interventions specific to A-T include therapeutic exercise, inspiratory muscle training, and early nutritional assessment and intervention. Further research will be required to determine the full potential of the identified interventions, including translatability to the A-T setting for evidence related to other forms of ataxia. Large gaps exist in the nursing and allied health evidence-base, highlighting a need for robust research that includes children and young people with A-T and their families to better inform and optimize management strategies.

Key-words ~~(4-6 from Index Medicus)~~: Ataxia telangiectasia,~~;~~ Cerebellar ataxia,~~;~~ Community health services,~~;~~ Child care,~~;~~ Rehabilitation,~~;~~ Multidisciplinary management

Introduction

Ataxia telangiectasia (A-T) is a rare genetic and progressive condition with prevalence estimates varying between 1 in 40,000 and 1 in 100,000 live births worldwide [1]. A-T primarily affects the neurological, immunological, and pulmonary systems, and typically first presents with cerebellar ataxia in early childhood. The neurological decline increases with age [64] and Pprogression is characterized by oculomotor, extrapyramidal, and peripheral nervous system symptoms, and most children are wheelchair-dependent by adolescence [1, 2]. Difficulty coordinating chewing and swallowing is common among children and young people with A-T, resulting in a high rate of malnutrition [3].

Lung disease is a significant manifestation of A-T and the leading cause of morbidity and mortality in the population [4, 5]. This results from weakness and incoordination of respiratory muscles and fibrosis of lung tissue [6]. Immunological deficiencies also drive a high malignancy rate in this population, in particular leukaemia and lymphoma, and result in the death of approximately 22% of people with A-T [5, 7].

People with A-T have complex needs and, in the absence of cure, require coordinated multidisciplinary care [2]. Appropriate symptom management and rehabilitation can improve their quality of life (QoL) and reduce the likelihood of complications [8] and it is essential that people with A-T have access to high-quality, collaborative care to minimize the burden of disease [9].

However, awareness of A-T is low, both in the clinic and community, and current guidance on the management of children and young people with A-T is generally limited to diagnosis and medical interventions [10]. Despite the tendency for patient care to be managed in the community setting, little attention has been given to the guidance of nursing and allied health professionals (AHPs), or sought to understand the views of children and young people and their families [11]. There is, therefore, a need to raise awareness of A-T within community

healthcare professions and to provide guidance specific to nursing and allied healthcare teams, to optimize the management of children and young people with A-T.

To address this need, a scoping review was undertaken to systematically identify and map the type, scope, and content of an extensive range of sources to compile expertise from nursing and AHPs on the care and management of children and young people with A-T.

Materials and Methods

A scoping review was selected as the most appropriate and inclusive means by which to map and synthesize material from a vast and diverse array of sources, addressing objectives to- (1-) fully determine the extent of the current literature, (2)- summarize the findings exploring narratively both the generalizsability and translatability of evidence to A-T, and (3)- identify research gaps [12].

This scoping review was performed according to the Joanna Briggs Institute methodology for scoping reviews [13], with reference to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines [14]. A protocol was published on an open science framework in August 2020 (https://osf.io/q7dba) and conducted using coproduction by the research team (https://osf.io/edzn3/).

Research Question and Outcomes

The research question was defined as follows: "*What guidance is available to inform the care and management of children and young people with A-T from the perspective of nursing and allied health professionals?*"

Key outcomes identified and mapped within the scoping review were as follows:

- Primary outcomes: Activity, participation, and health-related QoL.
- Secondary outcomes: Impairment level measures related to neuromusculoskeletal performance, cardiovascular performance, lung function, nutritional status, swallow, and ocular motor function.

Search Strategy

A comprehensive search strategy was developed in collaboration with an information retrieval specialist and clinical-academic experts in the selected fields using key words and medical subject headings (MeSH). Search terms (*Supplement 1*) were tailored according to the profession, with broader terms utilized where limited specific evidence was expected (for example, 'ataxia' was used for professions where a higher number of records were anticipated vs 'nervous system diseases' for those with fewer anticipated sources). A three-step search strategy was then undertaken as follows: (1). A search of PubMed and CINAHL electronic databases using the defined search criteria; (2). due to the high volume of initial results, remaining databases (*Supplement 2)* (including clinical trial and review registers, guidelines and recommendations, research libraries, and open grey literature sources) were searched using only the keyword 'ataxia'; (3). a manual search of the reference lists of all selected sources and consultation with A-Team experts from each discipline.

Nurses and AHPs representing the following therapists were selected for inclusion in this study, as those most likely to be involved in the care of people with A-T: physiotherapists (neurological and respiratory physiotherapy specialists), occupational therapists (OT), speech and language therapists (SLT), dietitians, orthoptists, optometrists, orthotists, and podiatrists. No restrictions were applied to date, language, location, culture, care setting, delivery type, or type of evidence and both progressive and non-progressive types of ataxia in children were included.

Screening and Selection

All sources of evidence were collated onto a web-tool called Rayyan QCRI [15] and their titles and abstracts were independently screened by two review authors (MK and EC). Evidence was selected that included ataxia in their title, population of ≤18 years of age with A-T or another ataxia impacting childhood. As there is limited evidence guiding the care and

management of A-T, other A-T-like conditions affecting childhood were included to ensure that all relevant knowledge with potential for translation to the A-T setting was captured. Care and management practices that focused on prevention, management or treatment of primary impairments, activity limitations, participation restrictions, or secondary health conditions, or interventions for health promotion, or provision of support and independence were considered. Surgical interventions were only included if nursing and/or any of the selected AHPs were significantly involved in pre- or post-surgical care.

Any material that did not meet the inclusion criteria, including any regarding medical, psychological, or pharmacological interventions, or evidence that did not consider management or intervention wereas excluded.

Full-text screening was conducted by two independent reviewers (MK and the discipline experts from the A-Team collaborative: EC, TP, LB, KD, NM, GQ, BC, SM, JP, HB, and RM). Reviewer discussion was conducted for sources of evidence where there was any doubt over inclusion, with a third review author (AW/LB) making the final decision where agreement was not reached.

Data Extraction

Information relating to the care and management of children and young people with A-T was extracted from the selected materials by the lead reviewer (MK) and verified by a second reviewer (discipline experts from the A-Team collaborative: EC, GQ, KD, TP, LB, BC, JP, and NM). Data were reported in a pilot-tested data-charting table adapted from the Joanna Briggs Institute reviewer's manual [12] to record citation details, context, intervention details, key findings and results, and direct generalizsability or transferability to A-T.

Results

A total of 21,118 sources of evidence were identified from database and grey literature searches of material related to allied health-care of A-T or similar conditions. After duplicate removal, the title and abstracts of 16,847 sources of evidence were screened, and 467 were selected for full-text screening. Of these, 79 could not be sourced, and 388 were screened. A total of 341 were excluded for the following reasons: (a) adult focus; (b) provided generic overview only, no intervention/therapy recommended; (c) pharmacological intervention; (d) focus on measurement or assessment of ataxia, no intervention/therapy recommended; and (e) standalone transcranial magnetic stimulation (TMS) treatment or variations. The reference lists of the remaining 47 sources were screened and three further studies meeting the selection criteria were included. This resulted in a final selection of 50 sources of evidence (Fig~~ure~~ 1).

Source Characteristics

Of the 50 selected sources of evidence, 42 were identified from PubMed and 8 came from CINAHL, Conference Proceedings Citation Index- Science (Web of Science), Cochrane Central Register of Controlled Trials, PROSPERO, and Google Scholar databases. A total of 35 were research studies (Table 1) and 15 were non-research material (Table 2, including reviews, book chapters, guidelines, and practice documents). Out of the 35 research studies, 31 were quantitative studies, 1 was a qualitative study, and 3 were mixed--method studies. Publication dates ranged from 1968 to 2022 with the highest number of selected evidence being published in 2017 (*n*=7) followed by 2021 (*n*=6). There were no orientating references to or information about the context in which the non-research sources of evidence were situated. The country of origin of the selected research studies included the USA (*n*=9), Brazil (*n*=6), Germany (*n*=4), UK (*n*=2), Italy (*n*=2), the Netherlands (*n*=2), Turkey (*n*=2),

Republic of Korea (*n*=2), Egypt (*n*=1), Iran (*n*=1), Australia (*n*=1), Israel (*n*=1), and New Zealand (*n*=1). One study represented multiple regions.

Of the AHPs included in the search strategy, physiotherapy was most heavily represented within the 35 research studies (*n*=23), followed by dietetics (*n*=7), speech and language therapy (*n*=2), respiratory physiotherapy (*n*=1), occupational therapy (*n*=1), and podiatry (*n*=1). No research studies related to nursing or optometry were identified. The non-research sources were mostly related to multiple allied health and nursing professions (*n*=9), while six were focussed specifically on physiotherapy or occupational therapy.

Of the 35 selected research studies with known intervention setting, the most commonly reported settings were hospitals/clinics (*n*=6), rehabilitation centres (*n*=6), A-T clinics (*n*=5), home (*n*=4), and laboratory/clinic and home (*n*=5).

The selected 35 research-based sources included more than 600 participants (where reported, female *n*= 304 and male *n*= 273) with ataxia as a primary or secondary impairment. The most number of participants included in a study w~~ere~~as 101 [44] and the least was one [16, 19, 21, 23, 24, 25, 28, 30, 53, 54, 60].) Where reported, the ages of children with ataxia ranged from 1.8 to 18 years (excluding the age of adult participants in mixed participant studies). Not all studies used functional descriptors to report severity of ataxia of their participants, but where reported, most participants were either wheelchair dependent or required assistance for mobility. Nineteen sources of evidence focused exclusively on children and young people with A-T (14 research and 5 non-research studies; reported age range in research studies was from 1.8- to 30 years). A-T--specific research studies related to dietetic (*n*=7), physiotherapy (*n*=3), SLT management (*n*=2), respiratory physiotherapy (*n*=1), and OT management (*n*=1). A-T-specific non-research studies generally provided multidisciplinary recommendations and practice advice, although none included advice on ophthalmic management despite the ocular motor impairment associated with A-T. Other progressive and inherited ataxias (Tables 1

~~&~~and 2) affecting childhood were the focus of 14 sources of evidence, while 14 focused on ataxia as part of a non-progressive condition. Finally, three sources reported mixed groups of participants with non-progressive and progressive ataxias (including A-T).

The sources of evidence reported a diverse array of care and management approaches which are reviewed here under five main themes of exercise and function, respiratory, nutrition, swallow, and speech. While exercise and function was the most researched theme with a total of 39 sources of evidence reporting findings/guidance on improvement of physical and functional performance~~,~~, the themes of respiratory and speech were least researched with only seven and six sources reporting evidence, respectively. Where reported, the duration of intervention ranged from ~~one~~ 1 day [22] to 125 weeks [20] and frequency ranged from one session per week [60] to three times daily [17]. The intensity ranged from 20 ~~minutes~~ [18, 41] to 120 min~~utes~~ per session (with 1~~-~~2~~-~~min~~utes~~ break in between sessions) [22]~~,~~, the most frequently cited duration being 60 min~~utes~~ (*n*=6).

Exercise and Function

Motor function decline is generally the first manifestation of A-T~~,~~ that increases with age [64]. ~~and t~~This was the most heavily represented theme in our search, with 37 sources of evidence reporting the impact of exercise and therapy on improving strength, coordination, and/or motor ability, and two sources of evidence reporting strategies to improve occupational performance. Although 12 studies examined the impact of physiotherapy, both the interventions and outcome measures varied considerably between studies [16-23, 53, 54, 57, 58]. Despite the wide range of outcome measures used, none of these studies with multiple participants~~,~~ focused on specifically evaluating age-dependent effects against treatment efficacy.

An A-T-specific case report published in 1968 emphasized the importance of prescribing a therapeutic exercise program, in particular following transition to a wheelchair [16]. This report highlights the value of passive to resistive exercises for retaining strength and range of motion, along with the importance of gait training, active assistive exercise, and wheelchair activities, together with the need for adaptive aids to facilitate children and young people's independence. However, the author has discouraged the use of overly demanding home programs to avoid adding to parental frustration and guilt [16]. Another A-T-specific case report [53] explored the effects of a 3-month-long physical therapy program including balance and strengthening exercises and balance games on a 9-year-old child with A-T. The study reported significant improvements in body structures and functions and positive improvements in the level of activity and participation following the therapy. Of the other ten studies reporting physiotherapy interventions, long-term multidimensional and individualized training programs generally led to improvements in strength, balance and stability, and/or functional parameters, which were often statistically and/or clinically significant although based on small sample sizes [17-23, 54, 57, 58].

The wide range of study types and methodologies utilized precluded any inter-trial comparisons or definitive conclusions. However, it is noteworthy that 6-months of intensive, goal-oriented functional rehabilitation intervals combined with vibration-assisted home training led to significantly improved motor function of children with ataxia [17]. Repeated practice of functional tasks led to improvement in balance and physical performance thereby improving overall motor function in children with cerebellar ataxia [54]. Reducing visual input during postural control [19], treadmill training in combination with transcranial direct current stimulation [18] or body weight support [23], standard physical therapy programs in combination with core stability program [58], and functional trunk training [57] were also

identified as physiotherapy interventions that may be worthy of further study in the A-T population.

Three case reports investigated the effect of assistive devices on gait performance, with mixed results [24, 25, 55]. Although robotic-assisted gait training (RAGT) led to decreased gait speed of a 16-year-old boy with non-progressive but persistent ataxia due to traumatic brain injury, the authors postulate that this may have been due to improved gait symmetry, decreased variability, and increased control achieved with a wearable robotic exoskeleton [24]. Conversely, RAGT in combination with conventional physical and occupational therapy led to improvements in postural control, functional mobility, and balance in children with ataxic cerebral palsy. Improvements observed after RAGT in areas of gross motor function that did not improve with conventional therapy alone support the potential benefits of over-ground RAGT on gross motor function in these children [55]. A conference proceeding from 2014 reported significant improvement from a dynamic movement orthosis suit and orthotic shoes in gait and stability for a 12-year-old girl with severe ataxia [25]. However, as these are case reports, the effectiveness of assistive devices for wider population remains uncertain and further research will be required to determine the potential benefit of such interventions for persons with A-T.

The benefits of tailored virtual reality and exergame-based coordinative training (including a combined approach with physiotherapy) were explored in six studies which mostly reported reduced ataxia symptomology following their respective intervention [26-30, 56], and in many cases this was considered to be clinically meaningful. Furthermore, the reported effects of immersive virtual reality and videogaming were considered to translate into daily living [26, 30]. Interestingly, two studies reported specific benefits of this approach on motivation

and goal attainment [26, 27]. One study also suggested that the degree of benefit was dependent on the amount of training, demonstrating the need for continuous and frequent training to optimize subsequent outcomes [26]. Another study that used exergames for upper body rehabilitation, reported improvement in hand dexterity of children with ataxia; the disease severity data, however, showed no significant change for participants [56]. Although A-T was only represented by three participants across all five studies [26, 30, 56], the positive outcomes following these interventions suggest they may warrant further study regarding their applicability to the management of children and young people with A-T.

The perspectives of parents and/or physiotherapists on the effects of physiotherapy were reported by three studies [37-39]. In an online survey of 96 physiotherapists [37] who treat children with ataxia, the most reported interventions were balance exercises (28%), task-specific training (23%), and proximal control activities (21%) and these were also ranked as the most effective types of treatment. Of the adjunct therapies reported, orthotics and walking/mobility aids (both 31%) were the most common and also ranked as the most effective. The common aims for physiotherapy treatment included improving coordination, balance, muscle strength, and educating the child/family regarding activity. The most used outcome measure was the Scale for the Assessment and Rating of Ataxia (SARA), followed by the Berg Balance Scale (BBS) and Gross Motor Function Measure (GMFM). However, no mention of any anticipated change in these measures or knowledge of minimal clinically important differences (MCID) related to physiotherapy intervention was provided by the respondents. This study provides a unique contribution to the understanding of good areas of practice, including multidisciplinary team rehabilitation and use of individualized treatment planning and standardized outcome measures from the perspective of physiotherapists [37].

In a study of parent perceptions of physiotherapy interventions for Friedreich ataxia, muscle strengthening and stretching were most reported, and the benefits of these were felt to increase as the child grew older. Despite this, parents generally perceived a home exercise program of muscle stretching to be challenging and burdensome, and reported adherence was low. Balance training was also commonly reported but only considered to be minimally helpful. Interestingly, hydrotherapy was the most recommended intervention by parents. Although parents reported a desire for more therapies, an array of barriers were identified to accessing services, both internal (including lack of time, understanding, or motivation) and external (including lack of access to professional services, lack of eligibility for services, and limited insurance coverage) [38]. Finally, a pilot study of parents who attended a training and support program in simple massage for use at home reported improved psychosocial well-being for both parents and children [39].

An occupational therapy intervention based on the Person-Environment-Occupation Model (PEO) was investigated on a child with A-T [60]. Based on the evaluation of the child's three aspects of occupational performance including the person, occupations, and environment, an individualised plan was designed to promote occupational performance. Intervention focused on the activities of daily living (such as dressing/undressing, toileting, eating/feeding), play, leisure, and social participation. The authors reported a significant improvement in the child's occupational performance and his participation in daily routines, enhanced self-confidence, and communicative skills.

Thirteen non-research sources provide guidance on the use of physiotherapy and exercise within a multidisciplinary approach for the management of A-T/ataxia [1, 2, 5, 31-36, 40, 51, 52, 63]. Physiotherapy and exercise were recommended as vital for maintaining strength,

coordination and balance, prevention of joint contractures, reduction of fatigue, and to maintain a person's independence for as long as possible, with the use of appropriate aids and devices (such as ankle-foot orthotics and weighted gait-aid) to maintain or improve daily activity [1, 2, 5, 31-35, 63]. Postural management was emphasized as being critical in maintaining functional ability, sitting
and standing balance, and respiratory function [5]. One systematic review identified a high number of interventions involving technology (video game or virtual reality) that have the benefits of being home-based which may facilitate beneficial outcomes [35].

Hydrotherapy, and even horseback therapy, were also recommended for children and young people with ataxia, with the added benefits of being well-tolerated and fun for them [1, 36]. Aquatic physical therapy and stretches were suggested to prolong ambulation and reduce the number of falls in people with Friedreich's ataxia [63]. The importance of early referral and initiation of physiotherapy, and for continued therapy, was highlighted [5, 31, 33].

The impact of the global COVID-19 pandemic on people with ataxia was also examined in a recent study. The report highlighted the importance of telehealth and telemedicine platforms in facilitating access to healthcare while reducing face-to-face visits. The study reported that online physiotherapy and balance therapy tutorials may support people in remaining physically active and engaged, prevent decompensation, address anxiety, and prepare them for the resumption of their normal routine [40]. Another recent editorial [62] that provided low-tech, inexpensive, and patient-centred advice on ataxia management, highlighted the impact of using face masks during COVID-19. Compromised visual feedback caused by the face mask restricted the field of vision and the additional effort of breathing through face mask impaired balance and reduced gait speed. Using visors instead of face masks, and

avoiding varifocal lenses offer some straightforward solutions. Some other suggestions to reduce the potential for tripping and falling included avoiding long clothes and footwear, wearing bags close to the body, discarding loose rugs/mats, and using weighed cutlery and chunky pens, among others.

Respiratory

Respiratory disease is a common and serious problem associated with A-T and was the subject of an A-T-specific longitudinal study that evaluated the effect of 24 weeks of inspiratory muscle training (IMT) on 11 children with A-T [41]. The intervention led to significantly improved ventilatory pattern, lung capacity, and respiratory muscle strength, alongside decreased respiratory rate. Significantly reduced dyspnoea and improved QoL (general health and vitality domains of the Short Form, SF-36) were also reported. The results suggest that IMT should be considered as an adjunct therapy to improve respiratory mechanics and QoL in children and young people with A-T [41]. This is supported by a case study of a child with A-T which emphasized the importance of postural drainage, coughing, and breathing exercises [16].

Respiratory management guidance was also identified in five non-research sources of evidence, with the common recommendations of improving pulmonary function and reducing aspiration within a multidisciplinary approach [1, 2, 5, 31, 42]. Guidance is also provided on removing bronchial secretions [1] and on regular airway clearance, including techniques to augment cough and mucociliary clearance [42], with an emphasis on regular activities and breathing exercises in optimizsing respiratory function [5].

Nutrition

Malnourishment was common among participants in the six studies that examined the impact of nutritional interventions in people with A-T [3, 43-47]. In each report, the study population tended to be of relatively low weight and height for age, with *Z*-scores appearing to decline with age [44, 45]. The progressive decline in nutritional intake led to recommendations for proactive consideration to be given to early intervention, in particular to percutaneous endoscopic gastrostomy (PEG) insertion at the start of BMI-Z decline, or from age 8 years onwards, to prevent progressive growth failure [44]. The limited available data suggest that early PEG placement may lead to improved weight gain [3, 44-46]. The benefit associated with earlier use of PEG as opposed to using PEG at the advanced stages of disease, was highlighted in a longitudinal study of the A-T population from Israel [45].

The poor oral intake, diet quality, and chronic fatigue experienced by people with A-T was described in a cross-sectional analysis that demonstrated the vulnerability of this population to issues influencing their nutritional status. The study further emphasizes the need for early nutritional intervention and ongoing nutrition support for families, including early discussions regarding tube feeding [3].

The identified non-research evidence in this theme also supports early nutritional intervention for people with A-T, including the management of pulmonary and nutritional complications of dysphagia, to improve clinical outcomes as well as positively impacting QoL [1, 2, 5, 31, 42]. Two sources of evidence also advocated for the involvement of a dietician to recommend dietary modifications [1, 5]. The importance of working closely with a dietician for dietary modification and dysphagia management is also highlighted to ensure optimal nutrition and hydration [5, 63]. A guidance document highlighted the importance of regular monitoring of weight and height to identify early signs of concerns and encouraged small frequent meals and snacks using nutrient-rich foods such as full-fat dairy products and food fortification [5]. A European consensus statement on the multidisciplinary respiratory management of A-T

also highlighted the nutrition- and swallow-related issues faced by this population, and the authors recommend assessment of the adequacy and safety of nutritional intake at least annually. They also recommend consideration of early PEG insertion for people with unsafe swallow or inadequate nutritional intake [42].

Finally, the issue of nutrition-related comorbidity was addressed by two research studies, one of which identified common metabolic disorders in people with A-T, specifically cardiovascular, diabetes, and liver diseases, and a tendency for these to worsen with age. The authors recommended nutritional and pharmacological interventions accordingly [43]. Elevated cardiovascular risk factors in the A-T population and the impact of poor nutrition and an altered lipid profile on the risk of developing atherosclerosis and diabetes were identified. The authors presented recommendations for routine monitoring of biomarkers and nutritional guidance [47]. Another cross-sectional study [59] reinforced the importance of assessing the nutritional status of selenium in the A-T population as they reported a significant inverse association between selenium concentrations (observed below the reference value in nearly 40% of A-T participants) and oxidative stress biomarkers.

Swallow

Oropharyngeal dysphagia with concomitant aspiration was the primary assessment made in a relatively large (n=70) study. Oropharyngeal dysphagia was commonly reported, and the authors concluded that it may be progressive in people with A-T. Aspiration was found to significantly correlate with lower weight/height Z-scores in people who presented with aspiration (27%). Aspiration also occurred with a higher frequency in young adults (with mean age 16.9 years) when drinking thin liquids through a straw, suggesting that this common feeding method may actually increase the risk of aspiration [46]. The authors

suggest videofluoroscopy as a technique to identify swallow dysfunction in young adults with A-T and recommend interventions to change the feeding routine to decrease the risk of aspiration and provide adequate nutrition and hydration. Such interventions include the elimination of thin liquids, pacing to slow the rate of liquid intake, and use of a feeding tube [46]. Other research studies that addressed the issue of swallow dysfunction identified the need for children to have their food mashed or cut into bite-sized portions to ease chewing difficulties, for thin liquids to be thickened to ease swallowing, use of a straw or sipper cup to reduce spillage, and for mealtime assistance, particularly in the evening when the child may be tired and slow to feed [3, 48].

These studies are supported by non-research-based sources of evidence, whereby a variety of recommendations were identified to support people with ataxia who experience difficulty swallowing [1, 2, 33, 63]. In addition to the interventions described above, education in safer practice at mealtimes and muscle strengthening to specifically target swallow pathophysiology were also recommended, with compensatory head postures and an appropriate sitting position considered helpful to facilitate safer swallowing [1, 2, 33, 63]. Other recommendations included environment modifications, such as reducing distractions and promoting focus on the task of swallowing by eliminating talking during meals; and dietary modifications such as altering the diet to exclude or modify textures and consistencies identified to cause coughing or choking. These may include dry crumbly foods (e.g.~~,~~ biscuits, nuts), small, easily inhaled foods (e.g.~~,~~ desiccated coconut, sesame seeds), and foods which are difficult to chew/breakdown (e.g.~~,~~ steak, apple) [63]. A multidisciplinary approach to dysphagia management was also recommended, incorporating the SLT, dietician, and physiotherapist/occupational therapist [33, 36].

Speech

Speech was addressed by a research study of eight children with A-T [48] which sought to determine deficits and assets in cognitive and speech-language functioning. The results revealed dysarthria in all participants and moderately-to-severely affected speech in most participants, with language functioning considered to be a strength. The authors emphasized the need for a valid assessment of cognitive and speech language functioning of the A-T population from an early age as a pre-requisite for providing appropriate support concerning the school environment. The authors suggested that children and their parents might be reassured that, although A-T is a severe neurodegenerative disorder, acquired intellectual and language skills are relatively well preserved [48]. However, expressive verbal and non-verbal language is significantly impaired in classical A-T in majority of children at school age [5]. Although more recent evidence on A-T suggests a cognitive decline with age affecting executive function, motor language function, spatial skills, affect, and social cognition [65, 66, 67], formal neurocognitive testing is hindered by motor and communication deficits [64, 65].

Another observational study investigated the characteristics and severity of dysarthria in children and adults with A-T [61]. The primary advice to improve speech included positioning the individual in a stable sitting position, attention to breath control and speaking at the beginning of an exhalation, and slowing down the rate of speech to improve speech intelligibility. The role of the SLT in facilitating communication skills and working jointly with a multidisciplinary team was also highlighted in three non-research-based sources of evidence [1, 33, 63]. These recommend that comprehensive assessment of a person's communication should take into account the impact of communication difficulties on daily living, and that alternative and augmentative means of communication should be considered [33, 63]. The importance of improving the underlying physiological support, managing communication environment, education on vocal hygiene and general vocal health, and the

interventions to improve the performance of listeners was also highlighted in a set of guidelines for the management of speech changes in FRDA [63].

Discussion

This is the first review undertaken to identify and map the literature regarding the care and management of children and young people with A-T by nurses and allied health professionals. In identifying and distilling the current literature, we have attempted to begin to address the knowledge gap surrounding the care of this population with the aim of providing an insight into current management practices and informing future research.

We identified a total of 50 sources of evidence that matched our search criteria from 21,118 search results. Three were RCTs, but the majority were case studies/reports and single case experimental designs (*n*=13), while within the 15 non-research sources, the majority were guidelines (*n*=4) and reviews (*n*=3). Physiotherapy interventions targeting impairments or activity limitations were most frequently identified (*n*=23/35 studies) with a diverse array of treatment types, intensity, frequency, and duration represented in both clinical and home-based settings.

Of particular note within the exercise-based research studies were six papers that examined the impact of technology. Although only three children with A-T were included within these studies [26-30, 56], the overall improvements observed in balance and ataxia highlight the potential for technology as a home-based rehabilitation strategy, reducing exposure to infections within acute healthcare settings and limiting travel-related fatigue. Hydrotherapy was also commonly recommended for A-T, including in combination with other exercises, and was the most frequent intervention recommended by parents for children with Friedreich ataxia. Although no A-T-specific research data were identified with regard to hydrotherapy, water is considered to reduce the effects of ataxia, making it easier to undertake physical exercise [1, 17, 20, 36, 38], and warrants investigation in the A-T setting.

Overall, all the studies that tested the effectiveness of interventions reported at least one positive outcome for the study population. However, despite change in scores, MCID were

largely not specified or reported upon, and sample size calculations were unreported, ultimately interpreted as statistically underpowered studies with limited generalis~~s~~zability to the A-T or wider ataxia populations. However, even though the study limitations presently preclude the direct generalization of results to the A-T population, these interventions may be worthy of future rigorously designed studies.

It is noteworthy that, within the limited evidence identified, discrepancies exist between the perspectives of physiotherapists and parents regarding the value of interventions, emphasizing the importance of gathering multiple viewpoints [37, 38]. We did not identify any studies that examined the perspective of the children or young people themselves regarding their health and well-being which presents a significant gap in the field. Although this is not uncommon [49], opportunities should be provided for children and young people to contribute their views about their healthcare and to steer research towards topics that are relevant to them and their everyday lives [50].

Finally, a common theme of the research- and non-research-based evidence relating to nutrition and dietetic interventions was for the early consideration of proactive PEG placement to prevent malnutrition [5, 44, 45]. The importance of early intervention was emphasized, with placement at advanced stages associated with poorer outcomes [1, 2, 5, 8, 31, 42]. As with each of the presented themes, management in this respect was recommended as part of a multidisciplinary team.

The methodology of this scoping review is limited by the capacity to only apply full comprehensive search strategies to 2 of the 13 databases, due to the high number of results obtained from the original search of PubMed and CINAHL. Our inability to source 79 clinical trial results from clinical trial database and conference proceedings registrations, despite our attempts to contact the corresponding authors, represents another major limitation

and we cannot rule out that relevant evidence may have been missed due to lack of reporting or publication bias.

This scoping review identified only a small number of research studies relating to the care and management of children and young people with A-T and indeed the wider populations of paediatric ataxia. The majority of these studies focused on physiotherapy and exercise-related interventions and no studies related to the care and management provided by nurses. A limited number of randomized controlled trials (*n*=3) highlights a considerable research gap in the field of A-T and paediatric ataxias. It could be argued that by the nature of A-T being a rare condition, RCTs are challenging to implement, and cohort studies may be viewed more favourably. However, guidance provided by less rigorous methods and even non-research sources of evidence still highlights gaps in knowledge available to guide nursing and AHP practices. Appropriately designed future studies are needed to provide reliable evidence on the effectiveness of interventions used in current practice.

Whilste the scoping review of the literature gives an overview of some aspects of multidisciplinary interventions, there are still many questions on what professionals would make the best team around the person with A-T, what criteria should trigger input from the different professions, and whether there are other groups (e.g., music or art therapists) not represented in the literature who could have an impact on the QoL of the A-T population. In summary, A-T is a complex multisystem condition and identifying a core set of standardized outcomes will be key to informing and advancing future practice and research. A multidisciplinary approach which includes children and young people with A-T and their parents will be necessary to ensure that all relevant outcomes are identified and prioritized.

Current research on A-T is limited. A-T research should be encouraged, and international, multi-centre collaborations will be required.

Conclusion

This scoping review provides the first comprehensive exploration of management approaches implemented by nurses and AHPs working with children and young people with A-T and other forms of childhood ataxia. A range of interventions were identified that are reported to positively impact ataxia-related impairments, activity, or participation levels, together with QoL measures. These findings are important when considering care and management strategies as they suggest there is the potential to improve outcomes for children and young people with A-T, or with similar conditions. Although limited, some of the evidence included within this review was specific to A-T and may, therefore, hold direct relevance to our research question~~;~~ most notably, therapeutic exercise, including coordinative training, IMT, and early nutritional intervention. However, this review makes evidence-guided rather than evidence-based recommendations for practice given the lack of rigorous research focused on children and young people with A-T. Further research will be required to fully determine the potential for non-A-T focussed evidence-based practice to successfully translate into the A-T population.

This scoping review has identified large gaps in the nursing and AHP evidence-base which indicate an urgent need for more robust research that include children and young people with A-T and their families. Whil~~st~~e fully powered studies may be challenging given the rarity of the condition, rigorous study design and transparent open-access reporting of anonymous trial data should be strongly considered by researchers to facilitate future systematic reviews with meta-analyses or meta-syntheses. Presently, a dearth of highly rigorous research in A-T appears insufficient to warrant systematic review but other research methods (such as Delphi

consensus involving international experts in the field of A-T) may be considered in the interim to guide contemporary practice.

Supplementary Information

Supplement 1: A detailed overview of search terms incorporated in the review

Supplement 2: A list of all the databases that were searched

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Author Contribution

Study conceptualization: The A-Team Collaborative (https://osf.io/edzn3/). Study design: M.K., L.B., E.C., T.P., and A.W. Reviewing: M.K., E.C., T.P., L.B., K.D., N.M., G.Q., B.C., S.M., J.P., H.B., R.M., and A.W. Data extraction: M.K. and verified by E.C., G.Q., K.D.,

T.P., L.B., B.C., J.P., and N.M. Analysis and interpretation: M.K. and E.C. Manuscript preparation: L.B. and M.K. Manuscript review: all authors.

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Availability of Data and Materials

All data generated or analyzed during this study are included in this published article or available as supplementary files.

Declarations

Ethical Approval~~:~~

Not applicable

Competing Interests~~:~~

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Table 1. Characteristics of selected research-based sources of evidence

S. no.	Author (year), country [citation]	Type of evidence source	Population(s)/ population size	Condition(s)	Allied health/ nursing profession	Themes	Intervention/ study assessment Training parameters (e.g. intensity, frequency, duration)	Reported key findings
1	Joveini et al. (2022) Iran [60]	Case report	CYP; *n*=1	A-T	Occupational therapy	Exercise and function	Occupational therapy intervention focusing on ADLs (such as dressing/undressing, toileting, eating/feeding), play, leisure and social participation (once a	**Significant improvement in child's occupational performance and his participation in daily routines,** enhanced self-confidence and communicative skills

							week for 10 weeks)	
2	Unes et al. (2021) Turkey [53]	Case report	CYP, *n*=1	A-T	Physiotherapy	Exercise and function	Balance and strengthening exercises, and balance games (3 days/week for 3 months)	Reported s**S**ignificant **improvements in body structures and functions** and positive improvements in the level of activity and participation following the therapy
3	Synofzik et al. (2013) Germany [30]	Rater-blinded intra-individual control	CYP, *n*=1	A-T	Physiotherapy	Exercise and function	Sequentially structured 12-week videogame-based coordinative training program	**Videogame-based coordinative training may benefit people with A-T, with effects translating into daily**

		study					(Nintendo Wii®)	**living.** SARA score decreased 4.4 points by end of training period (most pronounced for posture and gait), and improved sitting and stance
4	Arledge (1968) USA [16]	Descriptive case report	CYP, *n*=1	A-T	Physiotherapy	Exercise and function Respiratory	Case evaluation highlighting the role of PT	**Highlights the importance of a prescribed therapeutic exercise program**, including passive to resistive exercises, along with the use of adaptive materials. Importance of postural

								draining, coughing and breathing exercises
5	Romano et al. (2022) Italy [56]	RCT	CYP, *n*=18	Progressive and non-progressive ataxia (including A-T)	Physiotherapy	Exercise and function	Exergame-based exercise training (8 exercise sessions done 5 times/week for 12 weeks)	**Hand dexterity of participants improved in the intervention group,** and worsened in control group; no reported significant change in SARA score observed for IG but significant increment in score observed for CG
6	Yigit et al. (2022) Turkey [57]	RCT	CYP, *n*=20	Autosomal recessive hereditary ataxia	Physiotherapy	Exercise and function	Functional trunk training and trunk stabilization training (3 days/week of 8-	**Proposed training provided improvements in trunk control and**

				(including A-T)			week rehabilitation programme)	**upper limb functions.**
7	Barlow et al. (2007) UK [39]	Mixed method pilot study	Parents, *n*=24	Friedreich ataxia and A-T	Physiotherapy	Exercise and function	Impact of TSP for parents (eight ~~one~~1-h~~our~~-long weekly sessions)	**TSP in massage skills for use at home had reported psychosocial benefits for parents and children,** including significant improvements reported in parental anxiety ~~(P=0.011)~~, depression ~~(P=0.046)~~, perceived stress ~~(P=0.020)~~, general self-efficacy ~~(P=0.010)~~, life satisfaction ~~(P=0.045)~~

								and parental health status post participation in the programme. ~~(P=0.020)~~ Parents reported improvements in children's mobility, sleep patterns, energy and activity levels, relaxation, and happiness.
8	Elshafey et al. (2022) Egypt [58]	RCT	CYP; *n*=36	Cerebellar ataxic CP	Physiotherapy	Exercise and function	Core stability exercise program alongside physical therapy program (physiotherapy 3 times weekly,	**Statistically significant reduction** ~~**observed**~~**reported** **in the severity of ataxia and improvement in balance**, with stronger

							intervention group received additional core stability training for 30 min~~utes~~)	effects reported~~observed~~ for IG. Core stability training improved strength and endurance of muscles, trunk control, corrective postural control mechanisms, and ability to use the vestibular, somatosensory, and visual systems to maintain balance and stability
9	Lee et al. (2021)	Single□subject	CYP; *n*=1	Cerebellar ataxia	Physiotherapy	Exercise and	Problem-based task training consisting of	**Repeated practice of functional tasks led to**

	Republic of Korea [54]	experimental study				function	four walking tasks- a 10□step walk and return, walking while carrying an object, walking between parallel lines, and kicking a ball (10 mins/task, 8 sessions each in baseline and intervention phase followed by ~~one~~ 1-year follow-up. Total duration of intervention phase not reported)	**improvement in balance and physical performance** thereby improving overall motor function in children with cerebellar ataxia
10	Yoo et al.	Case	CYP; *n*=2	Ataxic CP	Physiotherapy	Exercise	Over-ground robot	**Improvements**

	(2021) Republic of Korea [55]	Report				and function	assisted gait training (Angel Legs M20) alongside conventional physical and occupational therapy (5 sessions/week for ~~one~~ 1 month)	**reported in postural control, functional mobility, and balance** in children with ataxic CP. Improvements observed after RAGT in areas of gross motor function that did not improve with conventional therapy alone
11	Maring et al. (2013) USA [38]	Descriptive mixed method study	Parents; *n*=30	Friedreich ataxia	Physiotherapy	Exercise and function	Semi-structured interviews of parents of children with FA to understand perceived	67% of children received PT intervention, most commonly muscle strengthening and

							effectiveness and barriers to PT	stretching (considered increasingly useful as the child gets older), following by balance training (minimally helpful). **Hydrotherapy was the most frequently recommended intervention by parents.** Muscle stretching HEP considered challenging and burdensome (9% reported consistent use).

								External barriers: lack of expert providers Internal barriers: Limited time and energy, lack of awareness and children's preference
12	Harris-Love et al. (2004) USA [21]	Case report	CYP; *n*=1	Friedreich ataxia	Physiotherapy	Exercise and function	"Enablement-disablement" process: long-term task-oriented bimanual reaching activities, functional strengthening, and gait training using a walker with tension-	Minimal changes reported in nine-hole peg test, single limb stance time and manual muscle testing. **69.4% decrease in gait speed** (42.9% on subsequent use of U-step walking stabilizer).

							controlled wheels and reverse-braking (12 months)	**43.7% increase in force variability. Reduced fall rate** (from 10 to 3 per month)
13	Grecco et al. (2017) Brazil [18]	Double-blind, sham-controlled, crossover, pilot study	CYP; *n*=6	Ataxic cerebral palsy	Physiotherapy	Exercise and function	Treadmill training plus anodal tDCS or sham tDCS (10 sessions over 2 weeks)	**Treadmill training led to improvements in functional balance and performance of mobility activities**. However, these effects were only maintained for ~~one~~1-month post-treatment when combined with tDCS

14	da Silva and Iwabe-Marchese (2015) Brazil [28]	Prospective, longitudinal and descriptive case study	CYP; *n*=1	Ataxic cerebral palsy	Physiotherapy	Exercise and function	Virtual reality and video game (Nintendo Wii®) balance training (40 sessions over 4 months) alongside kinesiotherapy	**Virtual reality may be beneficial when used in combination with kinesiotherapy.** Improved motor function (average GMFM-66 increased from 71.69 to 77.46) and balance (BBS increased from 48 to 53). No improvement was reported in gait parameters
15	Clopton et al. (2003)	Single-subject	CYP and adults; *n*=5	Cerebellar ataxia	Physiotherapy	Exercise and	Axial weight loading (10% of body	**Effect of axial weight loading was**

	USA [22]	design	(4/5 CYP)			function	weight) in four different conditions (reported in one session)	**inconsistent**, with gait worsening more often than improving
16	Cernak et al. (2008) USA [23]	Case report	CYP; *n*=1	Cerebellar ataxia following posterior fossa haemorrhage	Physiotherapy	Exercise and function	Intensive locomotor training with a BWS system on treadmill and ground for 5 days/week for 4 weeks (clinic) followed by 4 months of BWS (home)	**Locomotor training with BWS on a treadmill in combination with overground gait training may improve ambulatory function.** Significant functional gains: Gillette functional walking score improved from 2 to 6

								WeeFIM transfers and mobility subscale improved from 3 to 6 and 2 to 5, respectively. The number of unassisted steps improved from none to all after 6 months
17	Peri et al. (2019) Italy [29]	Longitudinal before and after pilot study	CYP; *n*=11	Ataxia secondary to ABI	Physiotherapy	Exercise and function	Tailored exergame training using GRAIL, an IVR integrated with a treadmill and motion capture system (20 sessions in ~~one~~ 1 month), plus	**Combined IVR and physiotherapy training may be an effective approach for ataxic gait rehabilitation.** Ataxia (walking endurance and balance)

							physiotherapy	reported to be significantly reduced after training: SARA: 10.5 to 8.5 ~~(P=0.012).~~ GMFM-88: 97.0 to 98.0 ~~(P=0.004)~~ BBS: 53.0 to 54.0 ~~(P=0.016).~~ Reduced gait variability reported
18	Anderl and Trammell (2017) USA [24]	Case study	CYP; *n*=1	Ataxia secondary to TBI	Physiotherapy	Exercise and function	Robotic-assisted gait training for 4 consecutive days plus traditional gait and balance training	**Robotic assisted gait training reduced gait speed** over 4 days which was retained 7 days post intervention

								(0.18 m/s slower than baseline), possibly due to improved step-time symmetry and control. No change in SARA score
19	Sartor-Glittenberg and Brickner (2014) USA [20]	Retrospective case series	CYP and adults; *n*=3 (data extracted/ reported for 1/3 CYP)	Severe cerebellar ataxia secondary to TBI	Physiotherapy	Exercise and function	Long-term multidimensional treatment program of individual and group therapy sessions to minimize ataxia and improve mobility, including balance, pool, rock, and multitasking	**Long-term multidimensional physical therapy is beneficial for people with ataxia.** Improved strength and body function. Balance improved 19 points (BBS 4 to 23) Improved performance

							climbing sessions	and with recommendations to ambulate with use of walker
20	Mulligan et al. (1999) New Zealand [19]	Within-subject study	CYP; *n*=1	Non-progressive congenital ataxia	Physiotherapy	Exercise and function	Comparison of two physiotherapy approaches (functional tasks with reduced visual input vs traditional trunk stability exercises)	**Reducing visual input during postural control exercises may be an effective approach.** Greater improvements in functional measures were reported following treatment with reduced visual input vs stability exercises; benefits were

								maintained to a greater extent with this approach
21	Schatton et al. (2017) Germany [26]	Rater-blinded, intra-individual control study	CYP and adults; *n*=10 (6/10 CYP)	Spinocerebellar ataxia	Physiotherapy	Exercise and function	12 weeks of coordinative home-based exergame training (Nintendo Wii® and Microsoft Xbox Kinect®) for trunk and postural control, individualized according to baseline scores	**Individualized training may be beneficial in progressive ataxia, with effects translating into daily living.** Meaningful 2.5 reduction of SARA score was reported~~; (P<0.01), which positively correlated on the amount of training~~

								~~(P=0.04) and was driven by posture and gait subscores (P=0.005).~~ Improvements reported in posture ~~(P<0.01)~~, gait ~~(P=0.005)~~, and higher-than-expected goal attainment in daily living
22	Hartley et al. (2019) Multiple countries [37]	Cross-sectional mixed method study	PT; *n*=96	Ataxia following surgical resection of posterior fossa	Physiotherapy	Exercise and function	E-survey of PT to determine international physiotherapy practice	**Most commonly reported interventions: balance exercises** (97%)**, gait re-education** (95%)**, proximal control**

				tumours				(93%)**; with balance exercises** (28%)**, task-specific training** (23%) **and proximal control activities** (21%) **used most often and ranked as the most effective**. Most commonly used adjuncts to therapy: mobility aids (31%) and orthotics (31%), and ranked as the most effective, followed by treadmill training. Access to virtual training facilities was

								reported by 57% of respondents, which was most commonly in children with posterior fossa tumours (73%). Key challenges reported: lack of resources, lack of evidence, impact of adjuvant oncology treatment and psychosocial
23	Martakis et al. (2019) Germany [17]	Retrospective study	CYP; *n*=45	Progressive and non-progressive ataxia	Physiotherapy	Exercise and function	Intensive, goal-oriented functional rehabilitation intervals, combined	**Intensive training, including vibration-assisted therapy, significantly improves**

							with vibration-assisted home training for 6 months	**motor function of people with ataxia.** Significant improvement in motor function was reported: Median GMFM-66 improved by 2.4 (non-progressive) and 2.9 (progressive) points. 1-MWT improved significantly in the non-progressive group. Improvements were preserved in people with progressive ataxia and could be further

								developed in people with non-progressive ataxia
24	Hon and Armento (2014) USA [25]	Case report	CYP; *n*=1	Ataxia, cerebral atrophy and hypopituitarism	Podiatry	Exercise and function	DMO suit	**DMO suit and sneakers led to significant improvement in gait,** with bilateral heel-strike and no near falls
25	Ilg et al. (2012) Germany[27]	Rater-blinded prospective cohort study	CYP; *n*=10 (7/10 CYP)	Progressive spinocerebellar ataxia	Physiotherapy	Exercise and function	8-week coordinative videogame-based training (Microsoft Xbox Kinect)	**Intensive coordination training with video games improved motor performance in participants with progressive cerebellar degeneration.**

								Ataxia symptoms reported to have significantly decreased~~:~~ ~~SARA: P=0.0078;~~ predominantly impacted by improved posture ~~(P=0.0003)~~ which correlated with training intensity~~.~~ ~~Dynamic gait index: P=0.04~~ a~~A~~longside improvements in gait and goal-directed leg placement. Participants reported

								the training as highly enjoyable and motivational
26	Felix et al. (2014) Brazil [41]	Longitudinal before and after study	CYP; *n*=11	A-T	Respiratory physiotherapy	Respiratory	IMT, 5 sessions/week for 24 weeks	**IMT may be an effective adjunct therapy for people with A-T.** IMT led to improved ventilatory pattern ~~(V_T; [P=0.015]~~; respiratory rate ~~[P=0.018])~~, lung volume ~~(Vc [P=0.002]~~, and respiratory muscle strength ~~(MIP [P<0.001]; MEP [P=0.001])~~.

								SSignificant improvements in QoL ~~(general health [P=0.009] and vitality [P=0.014] domains of SF-36)~~ and reduced dyspnoea ~~(P=0.022)~~ were also reported.
27	Andrade et al. (2021) Brazil [59]	Cross-sectional controlled study	CYP and adults; *n*=22	A-T	Dietetics	Nutrition	Assessment of selenium levels and relate them to oxidative stress and lipid status biomarkers in people with A-T	**Significant, inverse, and independent association reported between selenium concentrations and oxidative stress biomarkers.** Presence of selenium was

								below the reference value in nearly 40% and low Glutathione peroxidase (GPx) activity in the participants
28	Krauthammer et al. (2018) Israel[45]	Retrospective chart review	CYP; *n*=53	A-T	Dietetics	Nutrition	Assessment of long-term nutritional and gastrointestinal aspects	**Progressive growth failure and low nutritional intake was observed with age**; most prominently in people with cough and choking at mealtime. **Proactive insertion of a PEG should be considered once BMI-**

								Z begins to decrease. BMI-Z was inversely correlated with age ~~(P<0.004)~~, declining below minimal BMI percentiles after age 4 (boys) and 7 (girls). Relative calorific intake reduced with age ~~(P<0.002)~~ and correlated with BMI-Z ~~(P<0.05)~~. PEG in 12 people were associated with improved BMI-Z ~~(P<0.05)~~

29	Paulino et al. (2017) Brazil [43]	Cross-sectional control study	CYP and adults; *n*=18 (12 aged 5-–15 years, 6 aged 16-–25 years; this is a mixed sample of ages. Data from participants <18 is not identifiable)	A-T	Dietetics	Nutrition	Study evaluated metabolic alterations and liver involvement	**Metabolic disorders are observed in adolescent population with A-T and tend to worsen with age. Nutritional intervention and pharmacological intervention may be beneficial.** Malnutrition: 33.3% vs 5.9% in control group Metabolic alterations were common: Glucose metabolism alterations: 54.6%

								Hepatic steatosis: 64.7% AST:ALT ratio >1: 58.8% Dyslipidemia: 55.5% Insulin sum concentrations correlated positively with ALT ~~(P<0.004)~~ and age ~~(P=0.002)~~
30	Stewart et al. (2016) UK [44]	Prospective study, including nested case control study	CYP; *n*=101	A-T	Dietetics	Nutrition	Study evaluated growth over time, including an evaluation of 14 children with a PEG	**There is a decline in growth over time. PEG should be considered from age 8 onwards.** 35% of participants

								were recorded as underweight and 73% as short stature growth on ≥1 occasion. Weight, height and BMI *Z*-scores declined over time, most obviously after 8 years of age. There was a trend for improved weight in the 12 children with a PEG who had available data
31	Ross et al. (2015) Australia	Cross-sectional analysis	CYP; *n*=13 (1/13 adult)	A-T	Dietetics	Nutrition Swallow	Assessment of nutritional status	**Malnourishment is common in people with A-T. There is a**

	[3]							**need for early nutritional intervention.** 77% participants had short stature and 54% were underweight. Significant malnutrition was reported for 69% and this was observed to significantly increase with age ~~(P<0.001)~~. The majority of the participants (62%) had a poor diet, characterized by high fat and sugar. Key

								barriers to nutrition were chronic tiredness and the need for mealtime assistance
32	Andrade et al. (2015) Brazil[47]	Cross-sectional study	CYP and adults; *n*=13 (age range 4–24 years; average age 14.6. This is a mixed sample of ages. Data from participants <18 is not	A-T	Dietetics	Nutrition	Assessment of nutritional status, plasma concentration of vitamin E and markers of cardiovascular risk	**Lipid biomarker and vitamin E profiles require routine monitoring of cardiovascular biomarkers and nutritional guidance.** 30.8% of participants were malnourished and 23.1% had stunted growth. Median lean body mass index was

			identifiable)					significantly lower in the A-T vs control group ~~(P=0.003)~~. TG, CT, LDL-c, and non-HDL cholesterol concentrations were significantly higher and HDL-c concentrations were significantly lower vs healthy controls, as reported. Vitamin E: total lipid ratio was lower in people with A-T vs controls~~.~~
33	Lefton-	Observatio	CYP; *n*=70	A-T	Dietetics	Nutrition	Assessment of	**Oropharyngeal**

	Greif et al. (2000) USA [46]	nal, cross-sectional study				Swallow	oropharyngeal dysphagia with concomitant aspiration	**dysphagia is common and appears to be progressive in people with A-T.** Weight and height were abnormally low at all ages. Participants who aspirated were reported to ~~had~~ have significantly lower mean weight ~~(P<0.002)~~ and weight/height ~~*(P<0.001)*~~ *z*-scores than those who did not aspirate. 14 (27%) participants

								exhibited aspiration (10 exhibited silent aspiration); those who aspirated were reported to be significantly older than those who did not aspirate ~~(P=0.01)~~
34	Veenhuis et al. (2021) Netherlands [61]	Prospective observational cohort study	CYP and adults; *n*=22 (15/22 CYP)	A-T	Speech and language therapy	Speech	Paediatric Radboud Dysarthria Assessment (p-RDA) was used to evaluate dysarthria	**Primary advice to improve speech included positioning the individual in a stable sitting position, attention to breath control and speaking at the beginning of an exhalation, and**

								slowing down the rate of speech to improve speech intelligibility
35	Vinck et al. (2011) Netherlands [48]	Observational case series	CYP; *n*=8	A-T	Speech and language therapy	Speech Swallow	Evaluation of cognitive and speech-language function in relation to (oculo)motor function	**Decline in cognitive and language functioning appears to level-off; neuropsychological and language assessment should take A-T complexities into account.** All except the youngest participants had intellectual impairment (mild-to-

								moderate/severe). Cognitive deficits in attention, (non)verbal memory and verbal fluency were reported, alongside dysarthria and weak oral-motor performance. Chewing and swallowing ability appeared to decline with age. Language did not appear to deteriorate with age.

1MWT, 1-min~~ute~~ timed walk test; *ABI*, acquired brain injury; *A-T*, ataxia telangiectasia; *BBS*, Berg Balance Scale also known as the Functional Balance Scale; *BMI*, body mass index; *BWS*, body weight support; *CT*, total cholesterol; *CYP*, children and young people; *DMO*, dynamic

movement orthosis; *GMFF-66*, gross motor function measure-66; *GRAIL*, gait teal-time analysis interactive lab; *HEP*, home exercise program; *IMT*, inspiratory muscle training; *IVR*, immersive virtual reality; *MEP*, maximal expiratory pressure; *MIP*, maximal inspiratory pressure; *PEG*, percutaneous endoscopic gastrostomy; *PT*, physiotherapists; *QoL*, quality of life; *SARA*, scale for the assessment and rating of ataxia; *SF-36*, 36-item short form survey of quality of life; *TBI*, traumatic brain injury; *tDCS*, transcranial direct current stimulation; *TG*, triglycerides; *TSP*, training and support program; V_c, vital capacity; V_t, tidal volume

Table 2. Characteristics of selected non-research-based sources of evidence

	Author (year), [citation]	**Types of evidence source**	**Population(s)**	**Condition(s)**	**Themes**	**Key points**
1	van Os et al. (2017) [2]	Narrative guidelines	Not age specific	A-T	▪ Exercise and function ▪ Respiratory ▪ Nutrition	Recommendations for multidisciplinary treatment: ▪ Motor dysfunction should be treated with a multidisciplinary approach consisting of intensive support from a rehabilitation

					▪ Swallow	physician and AHPs ▪ Pulmonary management should include proactive monitoring and early intervention of pulmonary disease ▪ Nutritional and swallowing issues should be addressed early to promote growth, with interventions focusing on increasing oral intake/diet quality and preventing aspiration
2	Gatti and Perlman (2016) [31]	Review	Not age specific	A-T	▪ Exercise and Function ▪ Respiratory ▪ Nutrition	Recommendations for multidisciplinary treatment: ▪ Motor dysfunction management should involve supportive therapy, including early with continued physical therapy ▪ Pulmonary management should be multidisciplinary and include monitoring of recurrent infection, immune function,

						pulmonary function, swallowing and nutrition - Nutritional issues may benefit from early intervention
3	Rothblum-Oviatt et al. (2016) [1]	Overview	Not age specific	A-T	- Exercise and function - Respiratory - Nutrition - Swallow - Speech	- Motor dysfunction management should be multidisciplinary management (physical, occupational and speech therapies, alongside exercise) and may help maintain function but should not be used to the point of fatigue and should not interfere with activities of daily life. - Pulmonary issues should be managed with early intervention of respiratory symptoms. Annual pulmonary function testing should be performed in all children from 6 years of age - Nutritional and swallow issues may be addressed through education of safe practice

						and dietary modifications, with feeding tube recommended in appropriate circumstances
4	Bhatt et al. (2015) [42]	Review and practice document	Not age specific	A-T	Respiratory Nutrition	Statement on the multidisciplinary respiratory management of A-T, based on aggressive proactive monitoring and treatment of the various aspects of lung disease, including respiratory surveillance, regular assessment of lung function regular airway clearance, and input from speech and language therapist to address feeding issues
5	Taylor et al. (2014) [5]	Guidance document	CYP	A-T	Exercise and function Respiratory Nutrition	Guidance to professionals on the diagnosis and treatment of children with A-T. Main focus is on clinical care, but offers some guidance on therapeutic care and management
6	De Silva	Editorial	Not age specific	Ataxia	• Exercise and	Advice on low-tech, inexpensive, and patient-centred ataxia approaches for ataxia management.

	(2021) [62]				function	This included advice on resolving issues with gait and balance resulting from the use of face masks during COVID-19 pandemic; and reducing the potential for tripping and falling by attending to clothing and footwear, and making environmental adjustments
7	Hartley et al. (2019) [35]	Systematic review	CYP	Ataxia	Exercise and function	Promising results were reported from 11 studies (21 children with ataxia), but these were of low methodological quality and no firm conclusions were drawn regarding exercise and physical therapy
8	Woodford and Waterhouse (2021) [32]	Book chapter	Not age specific	Ataxia and Friedreich ataxia	Exercise and function	Handbook providing nursing guidance on the management of people with ataxia and Friedreich ataxia, with recommendations highlighting the need for multidisciplinary care

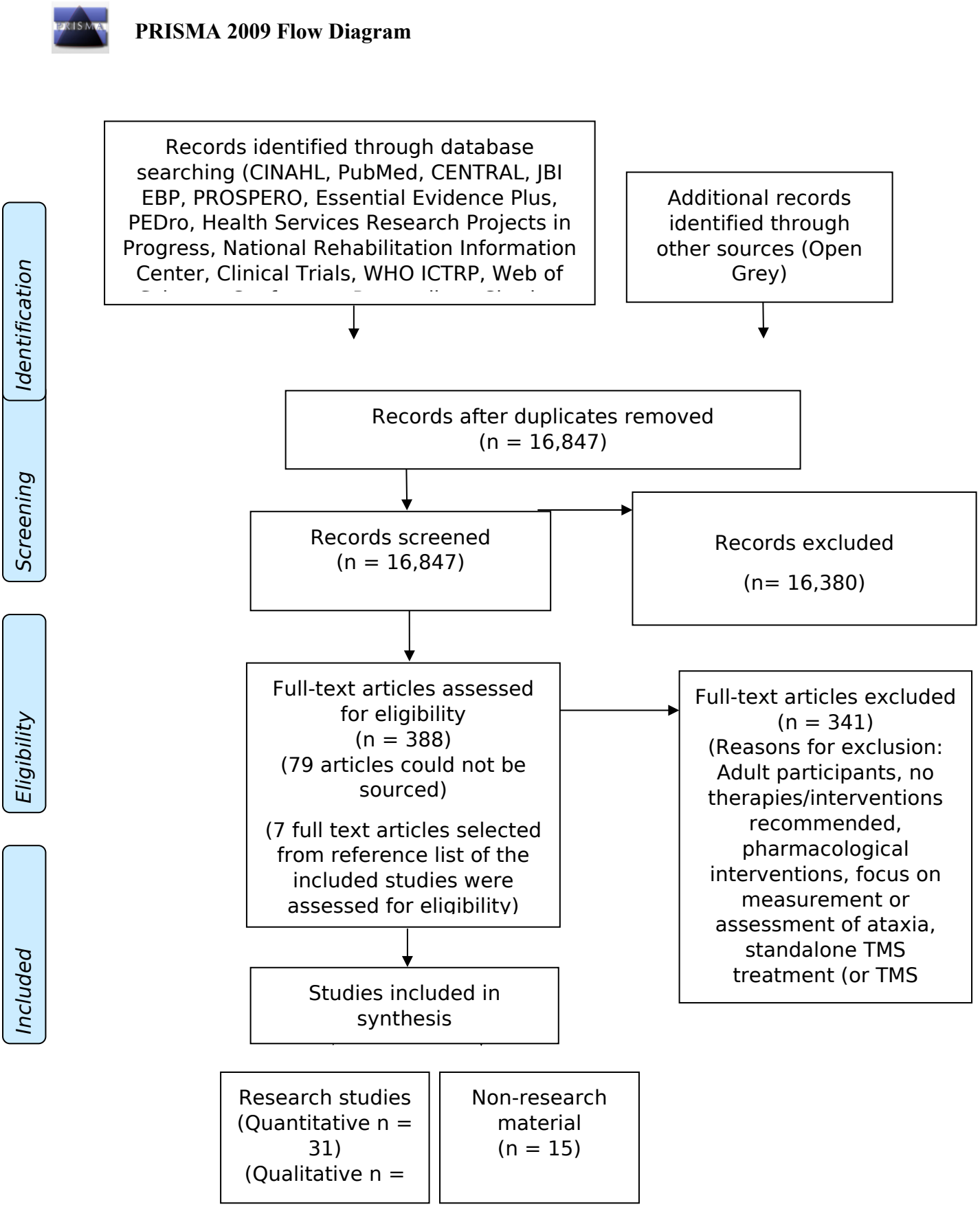
9	Maring and Croarkin (2007) [34]	Article	Not age specific	Friedreich ataxia	Exercise and function	Review highlighting the role of PT in implementing rehabilitation therapies focusing on strategies to maintain or improve participation in all contexts for as long as possible, as well as educating people with Friedreich ataxia and their families about the condition, potential therapeutic interventions, and realistic expectations regarding those interventions.
10	Manto et al. (2020)[40]	Practice document	Not age specific	Cerebellar ataxia	Exercise and function	COVID-19 Cerebellum Task Force consensus guidance on the management of cerebellar ataxia during the COVID-19 pandemic: Telehealth and telemedicine platforms are important to facilitating access to healthcare. Online physiotherapy and balance therapy tutorials help people remain physically active and engaged

11	Meneses et al. (2017)[51]	Case recommendations	CYP	Cerebellar ataxia	Exercise and function	Case report highlights the need for a holistic and structured assessment of needs, alongside long-term collaborations between clinicians and the family to facilitate shared -decision--making, with interventions adapting as needed
12	Tallaksen (2008) [36]	Overview	Not age specific	Hereditary ataxia	Exercise and function Swallow	This review of hereditary ataxia highlights the importance of appropriate and collaborative management for providing symptom relief and improvement of the prognosis
13	de Silva et al. (2019) [52]	Guidelines	Not age specific	Progressive ataxia	Exercise and function Speech Swallow	Guidelines on the management of progressive ataxia emphasize the critical role of AHPs, in particular physiotherapy, occupational therapy and speech and language therapy
14	Bates et al.	Guidelines	Not age specific	Progressive ataxia	Exercise and	These guidelines on management of progressive ataxia recommend that those affected should have

	(2016)[33]				function Swallow Speech	access to referral to a full range of therapies including speech and language therapy, physiotherapy and occupational therapy
15	Corben et al. (2014) [63]	Guidelines	Not age specific	Friedreich ataxia	Exercise and function Speech Swallow	The multidisciplinary guidelines emphasisszed the importance of physical therapy and exercise in improving balance, strength, flexibility and motor performance; and use of appropriate aids and devices to maintain or improve daily activity level Multidisciplinary advice on management of speech changes and swallowing difficulties in FRDA was given. Supplementary feeding techniques were also recommended to increase nutritional intake

A-T, ataxia telangiectasia; *CYP*, children and young people ≤18 years; *Adults*, people >18 years

Fig ~~ure~~ 1~~.~~ PRISMA flow diagram of the search results



~~Electronic Supplementary Material~~

~~Supplement 1: A detailed overview of search terms incorporated in the review~~

~~Supplement 2: A list of all the databases searched~~