European Journal of Paediatric Neurology 56 (2025) 24–37

Contents lists available at ScienceDirect

European Journal of Paediatric Neurology

journal homepage: www.journals.elsevier.com/european-journal-of-paediatric-neurology

Original article

Towards new perspectives: International consensus guidance on dystonia in pediatric palliative care☆

Anna Mercante [a,*], Nardo Nardocci [b], Emilio Fernández-Alvarez [c], Daniel E. Lumsden [d,e], Julie Hauer [f], Mercedes Bernadá [g], Ross Drake [h], Ulrika Kreicbergs [i,j], Rocío Palomo-Carrión [k], Marco Gemma [l], Philippe Coubes [m], Alfonso Fasano [n,o], Jean-Pierre Lin [d,p], Franca Benini [q], on behalf of the Pediatric Dystonia and Palliative Care Group and the European Paediatric Neurology Society (EPNS)

[a] *Department of Biomedical and Neuromotor Sciences (DIBINEM), University of Bologna, Bologna, Italy*
[b] *Dipartimento di Neuroscienze Pediatriche Fondazione IRCCS Istituto Neurologico "Carlo Besta", Milano, Italy*
[c] *Neuropediatric Department, Hospital, San Juan de Dios, Barcelona, Spain*
[d] *Complex Motor Disorder Service, Evelina London Children's Hospital, Guy's and St Thomas' NHS Foundation Trust, London, UK*
[e] *Research Department of Early Life Imaging, School of Biomedical Engineering and Imaging Sciences, King's College London, London, UK*
[f] *Division of General Pediatrics, Boston Children's Hospital, Boston, MA, USA*
[g] *Pereira Rossell Hospital Center and Asociación Española Pediatric Palliative Care Units, Montevideo, Uruguay*
[h] *Queensland Paediatric Palliative Care Service and Queensland Interdisciplinary Paediatric Persistent Pain Service, Queensland Children's Hospital, South Brisbane, Australia*
[i] *The Department of Women's and Children's Health, Paediatric Oncology and Haematology, Karolinska Institutet, Karolinska University Hospital, Astrid Lindgren Children's Hospital, Childhood Cancer Research Unit, Stockholm, Sweden*
[j] *Department of Population, Policy and Practice, Great Ormond Street Institute of Child Health, University College London, London, UK*
[k] *Nursing, Physiotherapy and Occupational Therapy Department. Faculty of Physiotherapy and Nursing, University of Castilla-La Mancha, Toledo, Spain*
[l] *Neuroanesthesia and Intensive Care Unit Fondazione IRCCS Istituto Neurologico Carlo Besta, Milano, Italy*
[m] *Unité Pathologies Cérébrales Résistantes et Unité de Recherche sur ses Comportements et Mouvements Anormaux (URCMA), Département de Neurochirurgie, CHU Montpellier, France*
[n] *Edmond J. Safra Program in Parkinson's Disease, Morton and Gloria Shulman Movement Disorders Clinic, Toronto Western Hospital, UHN. Division of Neurology, University of Toronto, Toronto, Ontario, Canada*
[o] *Krembil Brain Institute, Toronto, Ontario, Canada*
[p] *Children's Neurosciences, Department of Women and Children, Faculty of Life Sciences and Medicine, King's College London, London, UK*
[q] *Pediatric Palliative Care, Pain Service, Department of Women's and Children's Health, University Hospital of Padua, Padua, Italy*

ARTICLE INFO

Keywords:
Dystonia
Pediatric palliative care
Complex healthcare needs
International consensus
Delphi method

ABSTRACT

Background: Pediatric dystonias are associated with a broad spectrum of etiologies, resulting in a heterogeneous patient population in whom clinical presentation, evolution, and therapeutic needs may differ. These neurological symptoms are particularly common in children and adolescents with life-limiting and life-threatening conditions requiring pediatric palliative care (PPC). The impact on the child's quality of life is significant, as is distress for caregivers. Addressing and alleviating dystonia is key to providing good palliative care; however, there is limited evidence. A greater recognition and management of dystonia in this setting is urgently needed to provide appropriate interventions and care.
Objectives: To develop a standardized approach to dystonia in PPC.
Materials and methods: A two-round Delphi process explored the views of experts on the definition, assessment, monitoring, and treatment of dystonia in PPC. Professionals from different backgrounds and disciplines were invited worldwide. The final panel comprised 71 participants who completed a multi-statement online questionnaire.
Results: Fifty-three items were endorsed, providing expert, consensus-based recommendations.

☆ *Affirmation that all authors have read and complied with the Journal's Ethical Publication Guidelines* We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

* Corresponding author. Department of Biomedical and Neuromotor Sciences, via Massarenti 9 - Pad. 11, 40138, Bologna, Italy.
E-mail address: mercanteanna@outlook.com (A. Mercante).

https://doi.org/10.1016/j.ejpn.2025.04.003
Received 1 November 2024; Received in revised form 23 March 2025; Accepted 3 April 2025
Available online 10 April 2025

Conclusions: The limited clinical knowledge of childhood dystonia represents a challenge, especially in children with palliative care needs. This study is a first international consensus on dystonia in PPC and offers novel approaches to improving the dystonia-related burden and advancing clinical practice in this vulnerable population.

1. Introduction

Dystonia is defined by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures, or both [1]. This chronic, disabling symptom may occur in isolation or as one part of a more complex neurological presentation [2]. In addition to being one of the most common movement disorders [3], it is extremely heterogeneous in children and associated with numerous causes varying in severity, clinical course, and prognosis [4,5]. Pediatric dystonia frequently arises as a consequence of an acquired insult to the central nervous system [4,6,7], most commonly in cerebral palsy (CP) syndromes [8]. A generalized body distribution is typical [2], affecting global motor function and interfering with the child's daily activities and participation [3,9]. In many cases, dystonic symptoms pervasively progress often under-recognized [10]. Pain is an important component and cause of distress [11]. *Status dystonicus* is associated with potentially fatal complications [12,13].

In children with life-threatening or life-limiting conditions requiring pediatric palliative care (PPC), dystonia has been identified as a major contributor to the disease burden and decreased quality of life (QoL) [14,15]. As a source of lifelong disability, prompt identification and treatment of dystonia represent a significant healthcare and rehabilitation challenge [16,17]. However, pediatric guidelines are currently limited to CP [18]. The concomitant dearth of scientific evidence for this age group further complicates dystonia management, resulting in largely pragmatic therapeutic choices.

Given the lack of evidence-based guidance, a Delphi consensus was initiated with the aim of providing recommendations on dystonia in children with palliative care needs.

2. Materials and methods

2.1. Study design and participants

This work is part of a more extensive research program investigating the most troublesome symptoms of PPC that lack recommendations [15]. One companion study on sleep problems was recently published [19].

A two-round Delphi process was adopted to structure a multidisciplinary discussion among international experts. The goal was to determine the main concerns and priorities of dealing with dystonia in the PPC setting and reach a consensus on its recognition and management. The project workflow is summarized in Supplemental Fig. 1.

2.1.1. The Delphi method

This technique allows the generation of a reliable consensus opinion from a group of experts, anonymously measuring through an iterative and controlled feedback process their (dis)agreement on a topic for which evidence is limited [20]. It is widely used to evaluate current knowledge, identify future research areas, resolve controversies, and formulate recommendations and guidelines [21].

The Delphi method starts with formulating a preliminary set of statements by a Steering Committee (SC) knowledgeable on the subject [22]. A questionnaire is then developed and sent to a larger group of experts – the Delphi panel – to gather their opinions. Responses are analyzed to provide interim results demonstrating consensus and non-consensus areas [23]. Subsequent rounds include questionnaires containing non-consensus issues revised by the SC based on panel suggestions and continue until an acceptable level of consensus is achieved, no further consensus issues emerge, or the response rate is insufficient [24].

In the present study, the level of agreement/disagreement was measured using a 5-point Likert scale (1 = total disagreement; 5 = total agreement). Consensus was defined as $\geq$75% of panelists expressing a vote $\geq$4 (agreement) or $\leq$ 2 (disagreement). Voting was conducted online via SurveyMonkey. The whole process was supervised by two professional methodologists and one study facilitator.

2.1.2. Selection of the Steering Committee

The SC comprised 12 experts, six in pediatric dystonia and six in PPC, representing nine countries worldwide. Different pediatric disciplines and backgrounds were considered. Clinicians included an anesthesiologist, a neurosurgeon, neurologists, and palliative care physicians; a nurse and a physiotherapist also joined the project. SC members were recruited according to their significant contribution and expertise in pediatric dystonia and/or PPC, demonstrated by scientific publication, bibliometric impact, participation in congresses and seminars, advanced educational degrees, role/function, and years of experience. After reviewing the most relevant literature on the topic, the SC defined the core competency areas, drew up a list of statements to include in the online survey, and suggested possible members for the Delphi panel.

2.1.3. Set-up of the Delphi panel of experts

One-hundred and thirteen candidates were nominated from different disciplines, backgrounds, and geographic areas. Selection criteria included a minimum of five years of clinical experience and active scientific publications in pediatric movement disorders and/or PPC.

Each potential participant was sent an email invitation introducing the project. Those who accepted received a secure online link to access the survey platform.

2.2. Design and validation of the Delphi questionnaire

Multiple literature databases (PubMed, Google Scholar, Cochrane Library, Web of Science, and Scopus) were systematically reviewed according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines to collect existing literature on dystonia in PPC. Beginning with this literature review, the SC established three core areas of interest: i) Definition, ii) Assessment and Monitoring, and iii) Treatment. A designated member of the SC was appointed as the supervisor for each field of expertise: one for PPC and one for pediatric dystonia. The remaining members were responsible for drafting statements based on the core areas and their extensive clinical experience.

Eighty-one statements were proposed. The two supervisors subsequently selected the most relevant ones, which were then refined and collegially agreed upon for inclusion in the final Delphi survey.

2.3. Data collection and analysis

The online questionnaire contained a personal information sheet and a series of statements for panelists to evaluate anonymously and comment on. Voting instructions were sent by email, advising them to disregard any statements that fell outside their area of expertise. The first survey opened in September 2023, and the entire process was completed in March 2024.

Data were analyzed anonymously using descriptive statistics.

3. Results

The final Delphi panel, designated as the *Pediatric Dystonia and Palliative Care Group,* was composed of 71 experts from 19 countries worldwide, representing a diverse range of pediatric disciplines and expertise (Fig. 1). Palliative care physicians, neurologists, neurosurgeons, physiatrists, physiotherapists, and nurses were involved. Sixty-seven (94%) participated in the second Delphi round.

Consensus was reached on 53 statements, as detailed in Table 1. Tables 2, 3, and 4 provide an overview of the statements that were endorsed or not endorsed in each round, organized by specific areas of focus. Responders' rates varied for each statement, with a minimum of 63% and a maximum of 100% in Round 1, and a minimum of 70% and a maximum of 99% in Round 2.

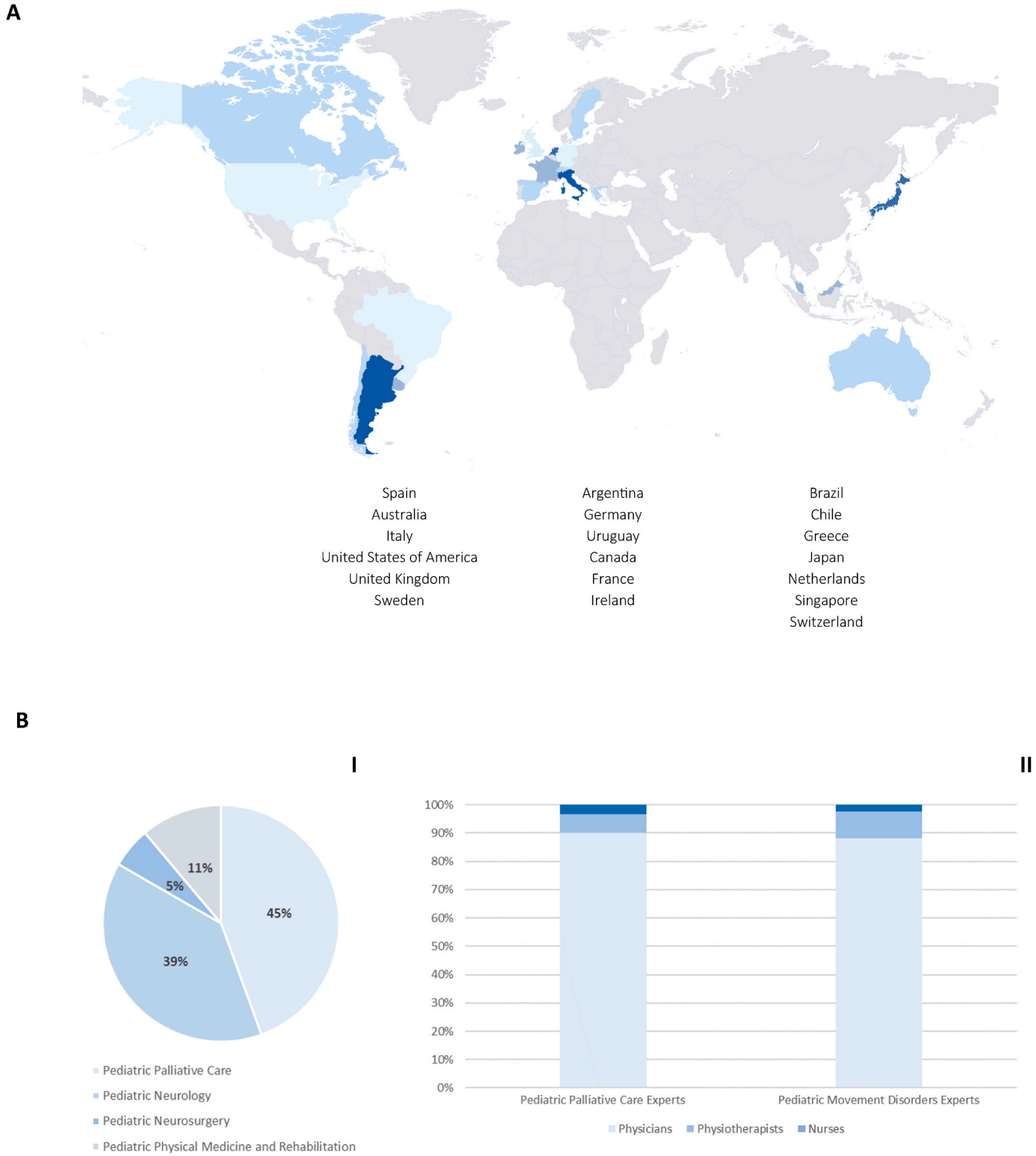

Fig. 1. Composition of the Delphi panel according to A. geographic location (in order of prevalence) and B. expertise (I) and discipline (II).

Table 1
Quantitative results of the two-round Delphi voting.

First Round	**n (%)**
Total consensus agreement	**28/60 (47)**
Definition	5/7 (71)
Assessment and Monitoring	5/13 (38)
Treatment	18/40 (45)
Second Round	
Total consensus agreement	**25/34 (73)**
Definition	1/1 (100)
Assessment and Monitoring	5/7 (71)
Treatment	19/26 (73)
Both rounds	
Total consensus agreement	**53/81* (65)**
Definition	6/7 (86)
Assessment and Monitoring	10/19 (53)
Treatment	37/55 (67)

* Of the statements not approved in Round 1, 19 were excluded and no longer considered, six were reformulated, six were split into two new statements, one into three, and 13 new statements were added.

3.1. First round

The Delphi process yielded 60 statements in Round 1: seven in Definition, 13 in Assessment and Monitoring, and 40 in Treatment. Twenty-eight of these obtained a positive consensus; specifically, 5/7 in Definition, 5/13 in Assessment and Monitoring, and 18/40 in Treatment.

3.2. Second round

Following panelists' suggestions, 19 of the statements not approved in Round 1 were excluded and thus no longer considered, six were reformulated, six were split into two new statements, one into three, and 13 new statements were added.

Thirty-four statements were voted on in Round 2 (one in Definition, seven in Assessment and Monitoring, and 26 in Treatment). A positive consensus was reached for 25 of them: 1/1 in Definition, 5/7 in Assessment and Monitoring, and 19/26 in Treatment.

4. Discussion

This study presents recommendations from the first expert consensus on dystonia in children with palliative care needs. We believe our results provide a framework to advance dystonia clinical practice and identify real-world criticalities and unmet needs, guiding future research.

Dystonia is a common movement disorder, yet concerns persist regarding its recognition [2]. Misdiagnosis and an underestimation of its prevalence are frequent [25,26]. The heterogeneity of PPC disorders that give rise to dystonia adds further clinical complexity. The panel agreed that consultation with a movement disorder specialist is recommended when dystonia is suspected (S9), and repeated longitudinal assessments rather than a single evaluation are advisable (S12). Early identification may be particularly challenging in the context of spasticity or other motor manifestations [27], making it difficult to assess the severity of the disorder and potentially delaying prompt targeted treatment when it is most efficacious [28,29]. Hence, accurate clinical observation and the ability to distinguish dystonia from other movement disorders are crucial [26].

Pain, dysautonomia, and bladder and respiratory dysfunction were recognized as non-motor manifestations during the vote (S4R). Although it is still debated whether they are strongly associated or secondary features of dystonia, their importance has been increasingly acknowledged [30,29]. Non-motor symptoms increase disease burden and disability while decreasing QoL [29]. Their systematic evaluation and management have already been suggested as routine clinical care [31]; however, no validated measures or recommendations on screening are available [29].

The panelists also strongly concurred on the importance of considering precipitants and chronic triggers (e.g., acute illness, acute and chronic pain sources, constipation, dysmenorrhea) in assessing and managing dystonia (S11), together with ongoing therapeutic interventions and disease progression (S13). In particular, it was deemed necessary first to exclude medication-induced dystonic reactions in new-onset dystonia (S22N). Other frequent triggers were identified in iatrogenic causes or related to arousal, cognitive tasks, and emotional status (S5).

Regarding assessment and rating tools, the panel concluded that the suitability of a particular dystonia scale in a PPC setting depends on several factors, including the characteristics of dystonia, the age of the child, and the established goals of assessment and care (S23N). However, existing scales present significant concerns in terms of reliability and content validity [32]. Most available evidence focuses on dystonia reduction or improvement in abnormal movements as a primary outcome, but these impairment-based scales fail to capture the complete

Table 2
Results of the Delphi survey on dystonia in PPC - Definition.

Statements		n	Likert scale Response (%)*					Consensus score, % (n)	Final result
			1	2	3	4	5	4+5	Approved
S1	The severity of dystonia often fluctuates with the posture assumed by the patient and voluntary muscular activation of the involved body area.	71	1	0	5	35	59	94 (67)	Yes
S2	Severe, generalized dystonia often combines with spasticity.	71	3	6	19	45	27	72 (51)	No
S3	In children with CP, dystonia appears as fluctuating hypertonia, involuntary postures, and abnormal movements.	70	0	3	6	37	54	91 (64)	Yes
S4	Associated features of dystonia include non-motor symptoms such as cognitive disability, behavioral disturbances, bladder dysfunction, and respiratory problems.	69	6	13	25	32	24	56 (39)	No
S4R	► Non-motor manifestations of dystonia include pain, dysautonomia, and bladder and respiratory dysfunction.	66	3	4	10	26	57	83 (55)	Yes, 2nd round
S5	Frequent trigger factors of dystonia are iatrogenic or related to arousal, cognitive tasks, and emotional status.	70	1	3	13	47	36	83 (58)	Yes
S6	Dystonia affects various aspects of quality of life, not limiting to those related to physical functioning.	71	0	6	0	4	90	94 (67)	Yes
S7	Dystonia generates significant distress in families of children receiving palliative care.	71	0	3	1	23	73	96 (68)	Yes

*Percentage results have been rounded up. Endorsed statements are reported in blue.
CP= cerebral palsy; n=number of voters; PPC= pediatric palliative care; S=statement.

benefits of interventions and the main concerns of children and their carers [5,30]. Our experts supported the implementation of indirect outcome measures that comprehensively address the contribution of dystonia to the child and family's global impairment and their functional priorities (S21N). Moreover, a consensus was reached to evaluate pain in all children with dystonia through appropriate and validated scales (S24N). Dystonia is frequently associated with chronic pain, which can intensify symptom burden and disability [8,11,33]. In some instances, pain has been found to impact QoL more than dystonia's motor severity [34,35]. Despite being a priority for many families dealing with dystonia [5,9,11], pain has received less attention there than in other movement disorders. Additionally, specific tools to assess dystonic pain are limited [33].

Dystonia in childhood is often unremitting and tends to worsen over time for most patients [10]. The panelists identified several valuable monitoring measures for worsening symptoms, including fragmented sleep, uncomfortable sitting, vegetative signs of stress and pain, and alterations in body temperature (S26N). These indirect parameters are of particular interest in settings of medical complexity. A considerable proportion of PPC patients with severe neurological impairment are unable to communicate, and may experience symptoms, including pain, differently [36].

The treatment of dystonia is primarily symptomatic involving medication, postural, surgical and rehabilitation interventions [37]. The current model of care is predominantly applied to adult-onset dystonia and centered on motor aspects [29]. However, the most common causes of dystonia differ between adults and children [38]. In some circumstances, specific treatments may be necessary [3,39], further limiting the application of the available general strategies and recommendations. Many therapies exist, but general agreement is often lacking [40], with approaches that may significantly differ between pediatric neurology and palliative care services [41].

The panel found a consensus on the need for a multidisciplinary and multifactorial approach to treating dystonia in PPC (S27) that should comprise non-pharmacological and pharmacological measures. Decisions regarding the best strategy for a particular child must be individualized according to the condition and the patient's and family's care goals (S67N).

It was also unanimously agreed that the treatment of exacerbating factors, including infection, constipation, gastrointestinal reflux, and pain, is a priority for managing dystonia (S70N).

Although several medications are used regularly as first-line management, evidence to support dystonia pharmacological agents in children is less consistent than in adulthood [3,30], with no licensed medication approved for the pediatric age [42].

In the present study, various statements aimed to distinguish drug options for chronic use from those limited to acute use. Medications put to the panel included anticholinergics, baclofen, benzodiazepines (BDZs), botulinum toxin (BoT), clonidine, dexmedetomidine, gabapentinoids, and levodopa (L-dopa). Interestingly, the degree of agreement regarding pharmacological treatment was relatively low. The panelists' different geographic proveniences might partially justify this, since medication availability and indications may change depending on the country of origin. Moreover, these differences most likely follow the lack of defined protocols for pediatric dystonia and possibly reflect differing clinical approaches and experiences between specialties, particularly regarding goals of care and co-existing problems that factor into drug selection.

The panel's endorsement of BDZs was limited to short-acting formulations as a feasible home rescue plan in case of exacerbations (S51R.1) and status dystonicus with the requirement of strict monitoring, given their possible association with adverse severe respiratory outcomes (S51R.3).

Furthermore, clonidine was recognized as a valid non-respiratory

Table 3
Results of the Delphi survey on dystonia in PPC – Assessment and Monitoring.

Statements		n	Likert scale Response (%)*					Consensus score, % (n)	Final result
			1	2	3	4	5	4+5	Approved
S8	Dystonia is a clinical diagnosis based on history, examination, and visual pattern recognition with no specific diagnostic tests.	70	0	6	10	31	53	84 (59)	Yes
S9	An evaluation by a movement disorder specialist is recommended to assess dystonia in the PPC setting.	69	2	1	17	28	52	80 (55)	Yes
S10	Creatine kinase (CK) is an essential biochemical marker in evaluating severe exacerbations of dystonia.	66	4	12	16	42	26	68 (45)	No
S10R	► In addition to the clinical evaluation, measuring creatine kinase (CK) when aligned with the goals of care may help identify unstable/worsening dystonia and adjust treatment.	64	0	8	30	30	32	62 (40)	No
S11	Assessment of precipitants and chronic triggers (e.g., drug toxicity or withdrawal, acute illness, acute and chronic pain sources, constipation, dysmenorrhea) is an integral part of the assessment and management of dystonia.	70	0	0	3	16	81	97 (68)	Yes
S12	Frequent re-evaluation of dystonia symptoms is required.	70	0	4	9	24	63	87 (61)	Yes
S13	The impact of any ongoing therapeutic intervention and the potential disease progression must be considered when evaluating dystonia.	70	0	0	0	24	76	100 (70)	Yes
S14	The Barry-Albright Dystonia Scale (BADS) is suitable for the PPC setting.	63	4	17	36	33	10	43 (27)	No
S15	The Unified Dystonia Rating Scale (UDRS) is suitable for the PPC setting.	61	3	18	51	21	7	28 (17)	No
S16	The Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) is suitable for the PPC setting.	63	5	16	47	27	5	32 (20)	No
S17	The Global Dystonia Rating Scale (GDS) is suitable for the PPC setting.	56	3	11	34	43	9	52 (29)	No
S18	The Dystonia Severity Action Plan (DSAP) is a suitable scale for the PPC setting.	60	3	2	27	48	20	68 (41)	No
S19	Dystonia Severity Assessment Plan (DSAP) helps monitor the effectiveness of management and progression to status dystonicus.	60	0	3	27	43	27	70 (42)	No
S20	Surface electromyography allows for identifying muscle activation patterns and better-addressing dystonia treatment.	67	10	24	45	16	5	21 (14)	No

S21N	Indirect outcome measures comprehensively addressing the contribution of dystonia to global impairment and functional priorities of children and their families must be implemented.	65	0	2	6	46	46	92 (60)	Yes, 2nd round
S22N	In children with new-onset dystonia symptoms, it is necessary first to exclude a dystonic medication reaction.	64	0	5	9	22	64	86 (55)	Yes, 2nd round
S23N	The suitability of a specific dystonia scale in the PPC setting depends on factors such as the etiology, type, and severity of dystonia, the age of the child, and the goals of assessment and care.	65	1	5	6	37	51	88 (57)	Yes, 2nd round
S24N	In all children with dystonia, assessing pain using appropriate and validated scales is crucial.	66	0	6	12	18	64	82 (54)	Yes, 2nd round
S25N	In all children with dystonia, assessing quality of life using appropriate and validated scales is crucial.	66	0	9	17	24	50	74 (49)	No
S26N	Fragmented sleep, uncomfortable sitting, vegetative symptoms of stress and pain, and alterations in body temperature are valuable indirect parameters to monitor during dystonia exacerbations.	64	0	0	8	41	51	92 (59)	Yes, 2nd round

*Percentage results have been rounded up. Endorsed statements are reported in blue.
n=number of voters; N= new; PPC= pediatric palliative care; S=statement.

depressant alternative to BDZs with a possible medication-sparing effect, particularly in patients with significant respiratory risk during exacerbations (S42R). In addition, it was considered a versatile therapeutic option for both basal and acute treatment in generalized dystonia, as it is rapidly titrated and administrable via different routes (S43R.1).

Interestingly, panelists considered neuropathic pain treatment (e.g., gabapentinoids) helpful in dystonia with pain unresponsive to first-line approaches (S55) and supported the use of gabapentin as additional medication, especially in severe dystonia when pain is a significant component (S73N). The possible integration of gabapentin with the routine management of pediatric dystonias is not new [30]. While its mechanisms of action are largely unknown [43], different studies have reported it as beneficial in relieving dystonia symptoms and improving pain and comfort [44,45].

Neurosurgical procedures and rehabilitation are known to play a significant role in dystonia [37]. Concerning surgical treatments, the panel supported the possible use of deep brain stimulation (DBS) in patients with documented drug resistance (S57) with structurally normal brain scans, or with significant disability and QoL decline regardless of age (S59). The potential for this therapeutic option requires a multidisciplinary selection process (S58R.1). In addition, intrathecal baclofen (ITB) was proposed as a beneficial therapy in cases of refractory dystonia and spasticity if aligned with the patient and family's goals of care (S72N). Interventions such as DBS have shown great potential for different dystonia syndromes [26], with a possible impact on the long-term outcome when treating patients earlier at the lowest efficient dose [46]. The recognition of the potential applicability of these surgical interventions in PPC, and within the appropriate caveats, has important implications for clinical practice, supporting the position that the diagnosis of a life-threatening or life-limiting condition should not preclude their consideration. Indeed, despite well-established measures for treating dystonia, neurosurgical approaches have sometimes been considered extreme and invasive in a palliative setting [38]. On the other hand, palliative care services are also responsible for providing chronic and not just end-of-life care, with some pediatric patients having long-life expectancies [47].

Another complexity of dystonia is the fluctuation of symptom severity over time, which at its most severe expression– *status dystonicus* - can be fatal [48]. The identification of the onset of this stage may be challenging [49]. The panel agreed that intensive care admission and early dependency should be considered when low-dose parenteral sedation is not possible and if aligned with the goals of care of the patient/family (S61R.1). In this perspective, "Advance Care Planning" was identified as an essential step in determining the place and intensity of dystonia care (S61R.2).

Despite being a cornerstone of chronic symptom treatment [37], scientific data supporting physical and other rehabilitative approaches remains poor [3]. However, the panelists were confident that physical therapy is a valuable tool for dystonia, optimizing functional abilities and mobility and reducing contractures or deformities (S62). Accordingly, the need for specialized training for physiotherapists and occupational therapists treating dystonia has been recently emphasized as a priority [40]. Gentle handling and manual techniques were also endorsed during the vote (S63), as well as devices and adaptive equipment such as splints, orthotics, or wheelchair modifications, which were recognized as potentially increasing comfort, mobility, and independence (S64).

Finally, a unanimous consensus was reached on the beneficial effect of psychological and emotional support for these children and their families (S65). Indeed, psychological support in the PPC setting encompasses all aspects of the patient's life and helps address disease-related challenges and identify strategies to cope with personal, relational, and social issues [47].

Several statements failed to meet a consensus.

Interestingly, none of the dystonia scales proposed in the study were endorsed for use in PPC (S14-S19). This could be related to the fact that numerous scales (with the exception of the Dystonia Severity Action Plan, DSAP) [50] are available to measure dystonia, but their application is often limited by being time-consuming, needing considerable training for use, having limited utility in severe cases, or presenting concerns in reliability, particularly for acquired dystonias, which are most common in childhood [32,51].

Again, creatine kinase (CK) failed to meet a consensus as a serum marker of severe dystonia exacerbations in PPC (S10R). If measuring CK

Table 4
Results of the Delphi survey on dystonia in PPC - Treatment.

Statements		n	Likert scale Response (%)*					Consensus score, % (n)	Final result
			1	2	3	4	5	4+5	Approved
S27	The management of dystonia requires a multidisciplinary and multifactorial approach.	69	0	0	0	14	86	100 (69)	Yes
S28	The main goals of the symptomatic management of dystonia in PPC include improving motor function, reducing pain, decreasing involuntary movements, and preventing articular contractures.	68	0	0	6	31	63	94 (64)	Yes
S29	Botulinum toxin is the first-choice treatment for pediatric focal dystonia.	67	3	15	12	46	24	70 (47)	No
S29R	▶ In children, botulinum toxin may be used to control disabling symptoms of focal or generalized dystonia.	60	0	5	18	35	42	77 (46)	Yes, 2nd round
S30	The administration of botulinum toxin correlates with the risk of cumulative dose that requires monitoring.	64	6	13	28	37	16	53 (34)	No
S30R	▶ The administration of botulinum toxin requires monitoring to avoid cumulative dose.	56	1	12	19	34	34	68 (38)	No
S31	Patients with cerebral palsy present more frequently adverse events to botulinum toxin.	65	11	31	43	9	6	15 (10)	No
S31R	▶ Patients with dyskinetic cerebral palsy require careful consideration before receiving botulinum toxin, being particularly at risk for adverse events.	58	0	15	33	29	23	52 (30)	No
S32	Ultrasound and electromyography-guided injections of botulinum toxin improve the targeting and the clinical outcome.	64	0	3	28	41	28	69 (44)	No
S33	In addition to motor problems, pain is a core parameter that guides the optimization of botulin toxin injection.	65	0	2	20	38	40	78 (51)	Yes
S34	Increasing doses and/or reducing the intervals between injections compensate for the poorer efficacy observed at the end of the botulin injection cycle, although they have a limited effect.	60	3	10	42	38	7	45 (27)	No
S35	Physiotherapy support to determine functional priorities is helpful before and following botulinum toxin injection.	66	0	0	5	30	65	95 (63)	Yes
S36	Levodopa is the first-choice treatment for pediatric generalized dystonia.	63	16	22	32	22	8	30 (19)	No

S36R.1	▶ When levodopa is indicated, a slow starting and titration of the dose is particularly recommended in this population.	57	0	2	10	39	49	88 (50)	Yes, 2nd round
S36R.2	▶ Even low doses of levodopa may produce marked choreic or dystonic movements; therefore, initial clinical monitoring is mandatory in all cases.	57	3	12	13	26	46	72 (41)	No
S37	The side effect profile of levodopa is usually favorable in children.	62	0	7	45	30	18	48 (30)	No
S38	Anticholinergics are advisable as second-line treatment in children with generalized dystonia after having excluded a response to Levodopa.	64	2	17	23	36	22	58 (37)	No
S38R.1	▶ Anticholinergics usually require prolonged treatment (weeks to months) before a clinical response is observed; therefore, adjusting the family's expectations is important to avoid a premature suspension.	60	0	2	11	39	48	87 (52)	Yes, 2nd round
S38R.2	▶ Acute titration and suspension of anticholinergics are contraindicated as they may be associated with exacerbations of dystonia symptoms.	59	0	8	11	47	34	81 (48)	Yes, 2nd round
S39	Anticholinergic medications must be started with low dosages and slowly increased as needed.	63	0	2	11	38	49	87 (55)	Yes
S40	Trihexyphenidyl usually requires prolonged treatment (weeks to months) before seeing a clinical response.	63	2	3	33	41	21	62 (39)	No
S41	Tetrabenazine adverse effects are common and most of them are dose-related.	61	0	5	46	38	11	49 (30)	No
S42	A chronic daily clonidine dose of 20 mcg/kg/day is indicated in pediatric dystonia.	61	13	21	35	23	8	31 (19)	No
S42R	▶ Clonidine is a non-respiratory-depressant alternative to benzodiazepines, particularly favorable in children with respiratory risk and in the acute setting where it may also spare the use of other medications.	63	0	3	18	28	51	79 (50)	Yes, 2nd round
S43	Administration of clonidine every 3 to 4 hours is useful for status dystonicus.	61	0	8	25	38	29	67 (41)	No
S43R.1	▶ In generalized dystonia, clonidine represents a versatile approach for both basal and acute settings as it can be rapidly titrated, administered via different routes, and converted to the transdermal patch.	62	0	3	13	36	48	84 (52)	Yes, 2nd round
S43R.2	▶ Continuous infusion of clonidine may help control the severity of status dystonicus if aligned with the goals and phase of care.	58	0	3	21	22	54	76 (44)	Yes, 2nd round
S44	The risk of clinically significant hypotension with clonidine is lower than the perceived risk.	64	0	8	22	51	19	70 (45)	No

S45	Baclofen is a more effective agent when dystonia is combined with spasticity.	65	3	0	20	55	22	77 (50)	Yes
S46	High doses of baclofen exacerbate axial hypotonia.	66	3	5	9	47	36	83 (55)	Yes
S47	The most common indication for ITB in children is spastic and dystonic cerebral palsy, in addition to metabolic and neurodegenerative conditions.	62	0	4	20	46	30	76 (47)	Yes
S48	Baclofen withdrawal occurs both with oral and intrathecal administration.	63	0	6	18	40	36	76 (48)	Yes
S49	Benzodiazepines are generally considered a second or third-line treatment for both focal and generalized dystonia.	64	3	9	18	54	16	70 (45)	No
S50	Benzodiazepines are well tolerated in the pediatric population and have fewer adverse effects compared with anticholinergics, baclofen, and levodopa.	64	5	33	37	22	3	25 (16)	No
S51	When treating dystonia, long-acting benzodiazepines are more effective compared with short-acting formulations.	63	0	13	28	45	14	59 (37)	No
S51R.1	▶ Short-acting benzodiazepines are a feasible home rescue plan in case of exacerbations.	63	0	0	18	35	47	82 (52)	Yes, 2nd round
S51R.2	▶ In small infants, long-acting benzodiazepines are a primary therapeutic option for basal therapy.	58	2	14	43	29	12	41 (24)	No
S51R.3	▶ Short-acting benzodiazepines may be helpful in acute dystonia and status dystonicus but demand cautious use and monitoring, given their possible association with adverse severe respiratory outcomes.	62	0	0	13	21	66	87 (54)	Yes, 2nd round
S52	In pediatric dystonia, a typical starting dose of clonazepam is 0.01-0.03 mg/k/day in two doses, with gradual titration to 0.1-0.2 mg/k/day in three doses.	62	2	3	27	52	16	68 (42)	No
S53	In pediatric dystonia, a typical starting dose of diazepam is 0.1 mg/k/day in two to four doses with gradual titration to the clinical effect.	63	5	2	30	52	11	63 (40)	No
S54	Intranasal dexmedetomidine is helpful in cases refractory to first or second-line treatment.	45	0	15	67	14	4	18 (8)	No
S54R.1	▶ Intranasal dexmedetomidine is a therapeutic option to consider in the acute care of refractory dystonia.	47	0	2	45	34	19	53 (25)	No
S54R.2	▶ Intravenous dexmedetomidine helps control the severity of status dystonicus if aligned with the goals of care.	52	0	0	33	27	40	67 (35)	No
S55	Neuropathic pain treatment (e.g., gabapentinoids) is helpful in dystonia with pain unresponsive to first-line treatment.	66	0	3	14	51	32	83 (55)	Yes
S56	Side effects of pharmacological approaches such as sedation are acceptable in refractory cases when improving dystonia symptoms control and QoL.	67	0	4	12	54	30	84 (56)	Yes
S57	DBS must be considered as a treatment option for patients with documented drug resistance.	63	1	3	18	35	43	78 (49)	Yes

S58	DBS has great potential for different dystonia syndromes in PPC prior to a well-selection of candidates.	59	2	17	25	24	32	56 (33)	No
S58R.1	▶ DBS has the potential for different dystonia syndromes as long as a multidisciplinary team properly selects candidates.	59	0	0	15	20	65	85 (50)	Yes, 2nd round
S58R.2	▶ In eligible early-onset dystonias, DBS should not be delayed because of age.	56	0	4	35	22	39	61 (34)	No
S59	Disability and reduced QoL are leading factors in considering DBS as a treatment option, independently of age.	66	0	5	16	45	34	79 (52)	Yes
S60	The benefit of DBS is maximum when dystonia is the main motor sign.	60	0	2	36	35	27	62 (37)	No
S61	If status dystonicus is not controlled with oral medications, more aggressive sedation and admission to ICU are mandatory.	65	3	11	15	17	54	71 (46)	No
S61R.1	▶ In status dystonicus, early high dependency or ICU admission should be considered in settings where low-dose parenteral sedation is not possible, depending on the goals of care of the patient/family.	64	0	0	9	27	64	91 (58)	Yes, 2nd round
S61R.2	▶ The place of care and intensity of the treatments must be part of the "Advance Care Planning" conversations with the patient/family and aligned with the goals of care in every phase of the disease.	66	0	0	6	8	86	94 (62)	Yes, 2nd round
S62	Physical therapy plays an essential role in the management of dystonia, optimizing functional abilities and mobility and reducing contractures or deformities.	67	0	0	5	34	61	95 (64)	Yes
S63	Gentle handling and manual techniques are helpful for dystonia symptoms in PPC.	66	0	3	9	39	49	88 (58)	Yes
S64	Devices and adaptive equipment such as splints, orthotics, or wheelchair modifications have the potential to enhance comfort, mobility, and independence in children with dystonia.	67	0	2	6	31	61	92 (62)	Yes
S65	Psychological and emotional support benefits both the child and their family members, helping to cope with emotional distress and promoting overall well-being.	67	0	0	0	20	80	100 (67)	Yes
S66	Caregivers' education and training on pediatric dystonia empower families to understand the condition, manage symptoms, and provide optimal care at home.	66	0	0	0	10	90	100 (66)	Yes
S67N	Therapeutic options must be individualized for each child, condition, and goal of care, with most children requiring a combination of treatments.	65	0	0	0	14	86	100 (65)	Yes, 2nd round
S68N	Reducing pain and global distress and increasing comfort are the primary therapeutic goals.	66	0	0	8	18	74	92 (61)	Yes, 2nd round

S69N	The impact of dystonia symptoms, their removal, and the weight of medication side effects on the child and their family (e.g., reducing mobility by reducing tone, urinary retention, constipation, and hypotonia) must be balanced in the treatment strategy.	66	0	0	0	20	80	100 (66)	Yes, 2nd round
S70N	When present, it is necessary to primarily manage exacerbating factors such as infection, constipation, gastrointestinal reflux, and pain.	66	0	0	0	20	80	100 (66)	Yes, 2nd round
S71N	Supportive treatment must include interventions to prevent or limit possible complications (e.g., muscle spasms, rhabdomyolysis) and the side effects of medications used for management, such as IV hydration, use of antipyretics and analgesics, promotion of comfort and sleep, monitoring of electrolytes and liver and renal function.	65	0	0	5	32	63	95 (62)	Yes, 2nd round
S72N	If aligned with the goals of care, ITB infusion may be beneficial for refractory dystonia in cases where spasticity coexists (i.e., cerebral palsy).	60	0	0	8	30	62	92 (55)	Yes, 2nd round
S73N	Gabapentin may be an effective additional medication to improve dystonic symptoms, especially in severe dystonia when pain is a significant component.	62	0	5	8	26	61	87 (54)	Yes, 2nd round

*Percentage results have been rounded up. Endorsed statements are reported in blue.
DBS= deep brain stimulation; ICU= intensive care unit; ITB= intrathecal baclofen; QoL= quality of life; n=number of voters; N= new; IV= intravenous; PPC= pediatric palliative care; S=statement

has been reported as helpful in identifying unstable/worsening dystonia and adjusting treatment to prevent metabolic decompensation and systemic complications [51,52], the context of PPC demands some precautions and non-invasive standards of care.

Concerning pharmacological treatments, the panelists did not agree on L-dopa for pediatric generalized dystonia (S36), although a trial is often recommended [3]. Also, they did not find any consensus on the applicability of dexmedetomidine in the acute management of dystonia, including *status dystonicus* (S54R.1, S54R.2). There is growing evidence regarding this clonidine-like adrenergic alpha-2 receptor agonist and its use in palliative care [53]. However, specific evidence on pediatric dystonia is limited to case reports [54,55].

In clinical practice, BDZs are a frequent second or third-line therapeutic option, with clonazepam and diazepam being two of the most common [52]. Studies comparing different BDZs in pediatric dystonia populations are few, but generally, longer-acting BDZs are adopted [3,56]. Here, no consensus was reached about the greater efficacy of long-acting BDZs compared to short-acting formulations (S51) nor on their dosages (S52, S53), or their use in small infants as first-line treatment (S51R.2).

Last, the panel did not concur on avoiding delays in starting DBS (S58R.2), contrary to some of the literature available [11,39]. Indeed, it has been suggested that a greater proportion of life lived with dystonia may reduce the neuromodulation efficacy of surgical interventions, in part because of the advent of progressive fixed deformities in children growing up with dystonia [51,57]. The question is whether this is amenable to change or is a marker of the poorer responsiveness of early-onset forms of dystonia compared to those that occur later in childhood. The fact that the above statement was not endorsed in our Delphi sessions likely reflects the dearth of evidence and experience of these neurosurgical approaches in PPC.

4.1. Future directions

Urgent means are required to identify diagnostic and management pathways for pediatric dystonia, with priority given to the definition of shared pharmacological approaches and lines of treatment.

Future work should focus on establishing a consensus set of dystonia-specific outcome measures meaningful to children with dystonia and their families/caregivers, valuing the impact of non-motor features.

Clinical aspects amenable to limiting the occurrence of acute exacerbations, progression to status dystonicus, or other chronic complications need major attention in this vulnerable patient group.

Biomarkers also remain under-explored. In patients with PPC needs, efforts are requested to identify and test feasible and reliable tools that help monitor dystonia and adjust treatment, when needed.

Lastly, new, clinically relevant studies should be performed to expand knowledge to help physicians, children, and their carers make appropriate decisions about neurosurgical interventions in pediatric dystonia and recognize their potential benefits.

4.2. Limitations of the study

This study has some limitations. While the Delphi process is methodologically powerful, it has been criticized for the low replicability of its results and for being influenced by the professionals involved. Although our study included participants with relevant expertise and was large enough to capture a range of perspectives, some similarities emerged in several statements. Additionally, some important topics relevant to clinical practice were overlooked, such as the timing for discontinuing dystonia treatment.

Moreover, our sample mainly represented developed countries. Clinical practices and research priorities in developing countries may differ and be poorly related to our results. Last but not least, the application of some recommendations may vary and be restricted by the PPC regional/national service structure and organization, as well as by

different policies regarding specific healthcare services, tools, and medications.

5. Conclusions

Dystonia is a common and impactful neurological symptom in children needing palliative care. Its identification and management represent a great challenge that needs global guidance. This international and multidisciplinary consensus aims to provide insights into the importance of dystonia in PPC and advance current clinical practice. These consensus-based recommendations are intended to develop a tailored model of care integrating evidence and expert opinion to possibly improve the quality of care and reduce distress among patients, caregivers, and healthcare providers. Although this is a preliminary study, the results obtained might help create the basis for developing guidelines and promote further studies focusing on the critical aspects and unmet needs identified here.

Declaration of patient consent

Informed patient consent was not necessary for this work.

Authors' roles

Study design: Franca Benini, Anna Mercante; data collection and interpretation: All; manuscript writing: Anna Mercante; manuscript editing: All; approval to submit: All.

Institutional review board or ethics committee that approved the study

Not applicable.

Funding sources

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Declaration of competing interest

None. The authors have no competing interests to declare relevant to this work.

Acknowledgments

Dr. Lin participated in the representation of the *European Paediatric Neurology Society (EPNS)* as co-chair of the *EPNS Movement Disorders Group*.

We express our deepest gratitude and appreciation to the Pediatric Dystonia and Palliative Care Group members who dedicated their time and efforts to participate in our study. Their valuable insights and input have significantly contributed to the success of this research project. In particular, we thank: Alcock Mark, Allen Nicholas M., Armitage Nicole, Assmann Birgit, Bergsträsser Eva, Bolognani Marco, Calvo Rocío, Carecchio Miryam, Carr Lucinda, Catalano Igor, Cerisola Alfredo, Chan Eunice K., Cif Laura, Cooper Monica S., Craig Finella, DiDomizio Galen, Farella Giuseppina Mariagrazia, Fernández-Medina Isabel María, Friedman Jennifer, Fuentes Leticia, Führer Monika, Garone Giacomo, Gelineau-Morel Rose, Gorman Kathleen M., Gorodetsky Carolina, Harvey Adrienne, Hoshino Kyoko, Hynson Jenny, Ibrahim George M., Katz Naomi, Kiman Rut, Koy Anne, Laddie Joanna, Landi Andrea, Lemmon Monica, Lumsden Daniel E., Madrid-Rodríguez Aurora, Marks Warren, Martín Sánchez María C., Martino-Alba Ricardo, Martinuzzi Andrea, Mherekumombe Martha, Mink Jonathan W., Mohammad Shekeeb, Moreno-Martínez Alejandro, Morgante Francesca, Nallar Martín, Ng Grace M.C., Nilsson Frans, O'Malley Jennifer, Peláez-Cantero Maria J., Pérez Ardanaz Bibiana, Phillips Marianne B., Piñero-Pinto Elena, Pons Roser, Puertas-Martín Verónica, Rey Andrea, Rodriguez Natalie, Romero-Galisteo Rita P., Roubertie Agathe, Rusalen Francesca, Sacks Bronwyn, Schteinschnaider Ángeles, Sejersen Thomas, Silveira-Moriyama Laura, Smith Martin, Soffritti Silvia, Stenmarker Margaretha, Treat Lauren, Wille Joakim, Willemsen Michael.

Support in data collection and analysis was provided by Simonetta Papa and Lara Vecchi, and proof editing by Michael John Hammersley; this assistance was covered by internal funds.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ejpn.2025.04.003.

References

[1] A. Albanese, K. Bhatia, S.B. Bressman, et al., Phenomenology and classification of dystonia: a consensus update, Mov. Disord. 28 (7) (2013) 863–873, https://doi.org/10.1002/mds.25475.
[2] Q.N. Luc, J. Querubin, Clinical management of dystonia in childhood, Pediatr. Drugs 19 (5) (2017) 447–461, https://doi.org/10.1007/s40272-017-0243-3.
[3] C. Gorodetsky, A. Fasano, Approach to the treatment of pediatric dystonia, Dystonia 1 (2022) 10287, https://doi.org/10.3389/dyst.2022.10287.
[4] A. Roubertie, F. Rivier, V. Humbertclaude, et al., [The varied etiologies of childhood-onset dystonia], Rev. Neurol. (Paris) 158 (4) (2002) 413–424.
[5] D.E. Lumsden, H. Gimeno, K. Tustin, M. Kaminska, J.P. Lin, Interventional studies in childhood dystonia do not address the concerns of children and their carers, Eur. J. Paediatr. Neurol. 19 (3) (2015) 327–336, https://doi.org/10.1016/j.ejpn.2015.01.003.
[6] V.H. Ganaraja, M. Netravathi, N. Kamble, et al., Spectrum of childhood dystonia evaluated at a tertiary care center from south India, Annals of Movement Disorders 4 (3) (2021) 136–142, https://doi.org/10.4103/AOMD.AOMD_8_21.
[7] V.N. Kumar, D. Garg, S.B. Mukherjee, S. Pandey, S. Sharma, Clinical and etiological profile of dystonia among children, Indian J. Pediatr. 90 (12) (2023) 1267, https://doi.org/10.1007/s12098-023-04719-1, 1267.
[8] H. Eggink, M.A. Coenen, R. De Jong, et al., Motor and non-motor determinants of health-related quality of life in young dystonia patients, Park. Relat. Disord. 58 (2019) 50–55, https://doi.org/10.1016/j.parkreldis.2018.08.008.
[9] H. Gimeno, A. Gordon, K. Tustin, J.P. Lin, Functional priorities in daily life for children and young people with dystonic movement disorders and their families, Eur. J. Paediatr. Neurol. 17 (2) (2013) 161–168, https://doi.org/10.1016/j.ejpn.2012.07.007.
[10] J.P. Lin, D.E. Lumsden, H. Gimeno, M. Kaminska, The impact and prognosis for dystonia in childhood including dystonic cerebral palsy: a clinical and demographic tertiary cohort study, J. Neurol. Neurosurg. Psychiatr. 85 (11) (2014) 1239–1244, https://doi.org/10.1136/jnnp-2013-307041.
[11] S. Perides, J.P. Lin, G. Lee, et al., Deep brain stimulation reduces pain in children with dystonia, including in dyskinetic cerebral palsy, Dev. Med. Child Neurol. 62 (8) (2020) 917–925, https://doi.org/10.1111/dmcn.14555.
[12] Vogt LM, Yang K, Tse G, et al. Recommendations for the management of initial and refractory pediatric status dystonicus. Mov. Disord.. Published online April 15, 2024:mds.29794. doi:10.1002/mds.29794.
[13] D.E. Lumsden, M.D. King, N.M. Allen, Status dystonicus in childhood, Curr. Opin. Pediatr. 29 (6) (2017) 674–682, https://doi.org/10.1097/MOP.0000000000000556.
[14] C.J. Wusthoff, R.A. Shellhaas, D.J. Licht, Management of common neurologic symptoms in pediatric palliative care: seizures, agitation, and spasticity, Pediatr. Clin. 54 (5) (2007) 709–733, https://doi.org/10.1016/j.pcl.2007.06.004.
[15] I. Avagnina, L. Giacomelli, A. Mercante, F. Benini, International project on troublesome symptoms in paediatric palliative care will focus on neuro-irritability, dystonia and sleep disorders, Acta Paediatr. 112 (3) (2023) 569–570, https://doi.org/10.1111/apa.16663.
[16] A. Roubertie, L.L. Mariani, E. Fernandez-Alvarez, D. Doummar, E. Roze, Treatment for dystonia in childhood, Eur. J. Neurol. 19 (10) (2012) 1292–1299, https://doi.

org/10.1111/j.1468-1331.2011.03649.x.
[17] L.M. Centen, D. Pinter, M.E. Van Egmond, et al., Dystonia management across Europe within ERN-RND: current state and future challenges, J. Neurol. 270 (2) (2023) 797–809, https://doi.org/10.1007/s00415-022-11412-4.
[18] D. Fehlings, B. Agnew, H. Gimeno, et al., Pharmacological and neurosurgical management of cerebral palsy and dystonia: clinical practice guideline update, Dev. Med. Child Neurol. 66 (9) (2024) 1133–1147, https://doi.org/10.1111/dmcn.15921.
[19] A. Mercante, J. Owens, O. Bruni, et al., International consensus on sleep problems in pediatric palliative care: paving the way, Sleep Med. 119 (2024) 574–583, https://doi.org/10.1016/j.sleep.2024.05.042.
[20] M. Niederberger, J. Spranger, Delphi technique in health sciences: a map, Front. Public Health 8 (2020) 457, https://doi.org/10.3389/fpubh.2020.00457.
[21] Z. Shang, Use of Delphi in health sciences research: a narrative review, Medicine 102 (7) (2023) e32829, https://doi.org/10.1097/MD.0000000000032829.
[22] S. Jünger, S.A. Payne, J. Brine, L. Radbruch, S.G. Brearley, Guidance on Conducting and REporting DElphi Studies (CREDES) in palliative care: recommendations based on a methodological systematic review, Palliat. Med. 31 (8) (2017) 684–706, https://doi.org/10.1177/0269216317690685.
[23] E. Taylor, We agree, don't we? The Delphi method for health environments research, HERD 13 (1) (2020) 11–23, https://doi.org/10.1177/1937586719887709.
[24] S. Chuenjitwongsa, How to: conduct a Delphi study, Medical Education Cardiff University, Published online, 2017.
[25] A. Albanese, F. Asmus, K.P. Bhatia, et al., EFNS guidelines on diagnosis and treatment of primary dystonias, Eur. J. Neurol. 18 (1) (2011) 5–18, https://doi.org/10.1111/j.1468-1331.2010.03042.x.
[26] A. Albanese, M. Di Giovanni, S. Lalli, Dystonia: diagnosis and management, Eur. J. Neurol. 26 (1) (2019) 5–17, https://doi.org/10.1111/ene.13762.
[27] H. Miao, A.M. Mathur, B.R. Aravamuthan, Spasticity and dystonia are underidentified in young children at high risk for cerebral palsy, J. Child Neurol. 37 (2) (2022) 105–111, https://doi.org/10.1177/08830738211059683.
[28] D. Graham, S.P. Paget, N. Wimalasundera, Current thinking in the health care management of children with cerebral palsy, Med. J. Aust. 210 (3) (2019) 129–135, https://doi.org/10.5694/mja2.12106.
[29] K.J. Peall, B.D. Berman, N. Bruggemann, et al., Non-motor symptoms in dystonia: from diagnosis to treatment, Dystonia 2 (2023) 11860, https://doi.org/10.3389/dyst.2023.11860.
[30] J.P. Lin, N. Nardocci, Recognizing the common origins of dystonia and the development of human movement: a manifesto of unmet needs in isolated childhood dystonias, Front. Neurol. 7 (2016), https://doi.org/10.3389/fneur.2016.00226.
[31] J. Junker, B.D. Berman, J. Hall, et al., Quality of life in isolated dystonia: non-motor manifestations matter, J. Neurol. Neurosurg. Psychiatry 92 (6) (2021) 622–628, https://doi.org/10.1136/jnnp-2020-325193.
[32] E. Monbaliu, E. Ortibus, F. Roelens, et al., Rating scales for dystonia in cerebral palsy: reliability and validity, Dev. Med. Child Neurol. 52 (6) (2010) 570–575, https://doi.org/10.1111/j.1469-8749.2009.03581.x.
[33] C. Listik, E. Listik, F. De Paiva Santos Rolim, et al., Development and validation of the dystonia-pain classification system: a multicenter study, Mov. Disord. 38 (7) (2023) 1163–1174, https://doi.org/10.1002/mds.29423.
[34] O. Kutvonen, P. Dastidar, T. Nurmikko, Pain in spasmodic torticollis, Pain 69 (3) (1997) 279–286, https://doi.org/10.1016/S0304-3959(96)03296-4.
[35] D. Page, A. Butler, M. Jahanshahi, Quality of life in focal, segmental, and generalized dystonia, Mov. Disord. 22 (3) (2007) 341–347, https://doi.org/10.1002/mds.21234.
[36] J. Hauer, Improving comfort in children with severe neurological impairment, Prog. Palliat. Care 20 (6) (2012) 349–356, https://doi.org/10.1179/1743291X12Y.0000000032.
[37] I.O. Bledsoe, A.C. Viser, M. San Luciano, Treatment of dystonia: medications, neurotoxins, neuromodulation, and rehabilitation, Neurotherapeutics 17 (4) (2020) 1622–1644, https://doi.org/10.1007/s13311-020-00944-0.
[38] L.A. Rasmussen, M.C. Grégoire, Challenging neurological symptoms in paediatric palliative care: an approach to symptom evaluation and management in children with neurological impairment, Paediatr. Child Health 20 (3) (2015) 159–165, https://doi.org/10.1093/pch/20.3.159.
[39] E. Fernández-Alvarez, Nardocci. Update on pediatric dystonias: etiology, epidemiology, and management, DNND 29 (April 2012), https://doi.org/10.2147/DNND.S16082. Published online.
[40] M. Smit, A. Albanese, M. Benson, et al., Dystonia management: what to expect from the future? The perspectives of patients and clinicians within DystoniaNet europe, Front. Neurol. 12 (2021) 646841, https://doi.org/10.3389/fneur.2021.646841.
[41] T. Slater, G. Hughes, D.E. Lumsden, J. Laddie, Management of dystonia in paediatric palliative care, Arch. Dis. Child. 104 (1) (2019) 102–103, https://doi.org/10.1136/archdischild-2018-315002.
[42] A. Koy, J.P. Lin, T.D. Sanger, W.A. Marks, J.W. Mink, L. Timmermann, Advances in management of movement disorders in children, Lancet Neurol. 15 (7) (2016) 719–735, https://doi.org/10.1016/S1474-4422(16)00132-0.
[43] G.J. Sills, The mechanisms of action of gabapentin and pregabalin, Curr. Opin. Pharmacol. 6 (1) (2006) 108–113, https://doi.org/10.1016/j.coph.2005.11.003.
[44] A. Harvey, M.C. Waugh, J. Rice, et al., A pilot feasibility study of gabapentin for managing pain in children with dystonic cerebral palsy, BMC Pediatr. 21 (1) (2021) 368, https://doi.org/10.1186/s12887-021-02847-1.
[45] N.Y.K. Liow, H. Gimeno, D.E. Lumsden, et al., Gabapentin can significantly improve dystonia severity and quality of life in children, Eur. J. Paediatr. Neurol. 20 (1) (2016) 100–107, https://doi.org/10.1016/j.ejpn.2015.09.007.
[46] L. Cif, P. Coubes, Historical developments in children's deep brain stimulation, Eur. J. Paediatr. Neurol. 21 (1) (2017) 109–117, https://doi.org/10.1016/j.ejpn.2016.08.010.
[47] F. Benini, M. Spizzichino, M. Trapanotto, A. Ferrante, Pediatric palliative care, Ital. J. Pediatr. 34 (1) (2008) 4, https://doi.org/10.1186/1824-7288-34-4.
[48] A. Iodice, F. Pisani, Status dystonicus: management and prevention in children at high risk, Acta Bio Medica Atenei Parmensis 90 (3) (2019) 207–212, https://doi.org/10.23750/abm.v90i3.7207.
[49] S. Followell, S. Amin, Dystonia: Identification, Investigation, and Management in Children, University Hospitals Bristol and Weston NHS Foundation Trust, Published online, 2023.
[50] D.E. Lumsden, C. Lundy, C Fairhurst, J.P. Lin, Dystonia Severity Action Plan: a simple grading system for medical severity of status dystonicus and life-threatening dystonia, Dev. Med. Child Neurol. 55 (7) (2013 Jul) 671–672, https://doi.org/10.1111/dmcn.12108.
[51] D.E. Lumsden, M. Kaminska, H. Gimeno, et al., Proportion of life lived with dystonia inversely correlates with response to pallidal deep brain stimulation in both primary and secondary childhood dystonia, Dev. Med. Child Neurol. 55 (6) (2013) 567–574, https://doi.org/10.1111/dmcn.12117.
[52] P. Termsarasab, S.J. Frucht, Dystonic storm: a practical clinical and video review, J Clin Mov Disord 4 (1) (2017) 10, https://doi.org/10.1186/s40734-017-0057-z.
[53] J. Gaertner, T. Fusi-Schmidhauser, Dexmedetomidine: a magic bullet on its way into palliative care-a narrative review and practice recommendations, Ann. Palliat. Med. 11 (4) (2022) 1491–1504, https://doi.org/10.21037/apm-21-1989.
[54] L. De Zen, S. Della Paolera, I. Del Rizzo, V. Taucar, A. Skabar, E. Barbi, Home intranasal dexmedetomidine for refractory dystonia in pediatric palliative care, J. Pain Symptom Manag. 59 (6) (2020) e3–e5, https://doi.org/10.1016/j.jpainsymman.2020.02.022.
[55] L. De Zen, A. Divisic, G. Molinaro, S. Solidoro, E. Barbi, Dexmedetomidine at home for intractable dystonia and insomnia in children with special needs: a case series, J. Pain Symptom Manag. 66 (6) (2023) e653–e657, https://doi.org/10.1016/j.jpainsymman.2023.07.018.
[56] G. Garone, F. Graziola, F. Nicita, et al., Prestatus and status dystonicus in children and adolescents, Dev. Med. Child Neurol. 62 (6) (2020) 742–749, https://doi.org/10.1111/dmcn.14425.
[57] D.E. Lumsden, H. Gimeno, M. Elze, et al., Progression to musculoskeletal deformity in childhood dystonia, Eur. J. Paediatr. Neurol. 20 (3) (2016 May) 339–345, https://doi.org/10.1016/j.ejpn.2016.02.006.