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2022-04

Markovic , I P , Nikolic , D , Stahl , M , Tederko , P , Hdyrya , O , Negrini , S , Zampolini , M & Kiekens , C 2022 , ' Evidence-based position paper of the UEMS PR M on the role of Physical and Rehabilitation Medicine (PR M) physician in the management of children and adults with spinal dysraphism ' , European Journal of Physical and Rehabilitation Medicine , vol. 58 , no. 4 , pp. 511-519 . https://doi.org/10.23736/S1973-9087.22.07536-0

http://hdl.handle.net/10138/352112
https://doi.org/10.23736/S1973-9087.22.07536-0

European Journal of Physical and Rehabilitation Medicine 2022 August;58(4):511-9
DOI: 10.23736/S1973-9087.22.07536-0

GUIDELINES

Evidence-based position paper of the UEMS PRM on the role of Physical and Rehabilitation Medicine (PRM) physician in the management of children and adults with spinal dysraphism

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ABSTRACT

Spinal dysraphism (SD) or spina bifida (SB) is a congenital deformity that results from embryonic neural tube closure failure during fetal development. This evidence-based position paper represents the official position of the European Union through the UEMS PRM Section. This paper aims to evaluate the role of the physical and rehabilitation medicine (PRM) physician and PRM practice for children and adults with spinal dysraphism. A systematic literature review and a consensus procedure involved all European countries delegates represented in the UEMS PRM section through a Delphi process. The systematic literature review is reported together with thirty-two recommendations resulting from the Delphi procedure. The professional role of the PRM physician requires specific expertise in the treatment of patients with SD to plan, lead and monitor the rehabilitation process in an interdisciplinary setting and to participate in the assessment of the needs of these patients in the transitional phase from childhood to adulthood, with particular attention to the activity limitation and participation restriction.

(*Cite this article as:* Petronic Markovic I, Nikolic D, Stahl M, Tederko P, Hdyrya O, Negrini S, *et al.* Evidence-based position paper of the UEMS PRM on the role of Physical and Rehabilitation Medicine (PRM) physician in the management of children and adults with spinal dysraphism. Eur J Phys Rehabil Med 2022;58:511-9. DOI: 10.23736/S1973-9087.22.07536-0)

KEY WORDS: Spinal dysraphism; Rehabilitation; Physical and rehabilitation medicine; Guideline Adherence; Consensus.

Introduction

Neural tube defects (NTD) are congenital anomalies that result from the abnormal embryonic development of the fetal nervous system. It is considered that NTD development is multifactorial in its origin, with both genetic and non-genetic factors playing certain roles.[1] The NTD prevalence in Europe and the Middle East is 1 per 1000 live births. Two main entities of NTD are spinal dysraphism (SD) and anencephaly,[1] with hydrocephalus as a common complication.

SD or spina bifida (SB) is a congenital deformity that results from embryonic neural tube closure failure during fetal development, at approximately 28 days (3-4 weeks) of gestation.[2, 3] The occurrence of SD is approximately 2.7 to 3.8 per 10,000 live births.[4] Furthermore, 90-95% of those with SD are with no previous positive family history.[3]

With the advancement in medical and surgical treatments of SD, there is an increase of adults with such conditions.[5] Le and Mukherjee stressed that more than 85% of children with SD survive into adulthood.[5] Increased surviving rates in this population of patients, lead to the rise of new challenges in diagnostic procedures, treatment, management, inclusion, and follow-up of these individuals.

In patients with SD, numerous complications were described, such as neurological, urological, gastrointestinal, orthopedic, musculoskeletal and dermatological.[3, 6-9] Moreover, it was stated that SD remains a significant cause of chronic disability for affected individuals.[9] Spinal deformities, including kyphosis and scoliosis, as well as lower extremities deformities including dislocations and subluxations of the hip and foot deformities, are common orthopedic and musculoskeletal conditions in patients with SD.[7] Thus, patients with SD, particularly children, could be challenged in physical functioning and their developmental process, especially in the early period of motor development. Motor and sensory dysfunctions associated with the neurogenic lesions in patients with SD might affect mobility.[10] Furthermore, bowel and bladder dysfunctions were noted in approximately 76% of both children and adults with SD.[8] All these issues are associated to different degrees with limitations in both activities (mobility, self-care, cognitive function) and participation (study, employment and social integration) of these patients.[9]

The needs of this paper came from the absence of uniform criteria in Europe for diagnostic evaluation (clinical, electromyoneurography (EMNG), urodynamics, imaging studies, genetic, functional assessment, etc), for the definition and implementation of rehabilitation programs (duration, modes, interventions, etc). Furthermore, there is a grey zone concerning the role of physical and rehabilitation medicine (PRM) in diagnostics, treatment and follow-up (short and long term) for these patients. Additional aspects such as heterogeneous pathology and a wide area of diagnostics and treatment (multi-, inter- and transdisciplinary) were also important.

This paper aims to evaluate the role of the PRM physician and PRM practice for children and adults with spinal dysraphism.

Methodology

This evidence-based position paper (EBPP) is drafted and produced according to the methodology proposed by the professional practice committee of the PRM section of the Union of European Medical Specialists (UEMS PRM Section).[11] It comprises two parts: "Systematic review of the literature" and "consensus with Delphi procedure among UEMS PRM section delegates."

Literature search

The systematic review of the literature was performed considering the professional relevance for PRM physicians and was judged by two authors of the paper (IP and DN). The search on SD was conducted with specified terms/strings and included articles extracted from PubMed/MEDLINE. The main inclusion criteria were the relevance of articles/reviews for the PRM profession. Additionally, grey literature was searched for the guidelines that are not published in Journals (online sources), including representative papers by relevant international bodies (*i.e.* UEMS PRM section). The period of searched articles was between 2015-2020 years. Our priority in systematic literature search was: systematic reviews, meta-analyses, randomized controlled trials, evidence-based papers and guidelines. Moreover, other relevant papers were considered as well.

Search strings in PubMed/MEDLINE: ((spinal dysraphism)[All fields] OR (spina bifida)[All fields]) AND (rehabilitation)[All fields] (String 1) #61; ((spinal dysraphism)[All fields] OR (spina bifida)[All fields]) AND (physical therapy)[All fields] (String 2) #31; ((spinal dysraphism)[All fields] OR (spina bifida)[All fields]) AND (exercise)[All fields] (String 3) #12; ((spinal dysraphism)[All fields] OR (spina bifida)[All fields]) AND (treatment)[All fields] (String 4) #268; ((spinal dysraphism)[All fields] OR (spina bifida)[All fields]) AND (diagnostics)[All fields] (String 5) #212; ((spinal dysraphism)[All fields] OR (spina bifida)[All fields]) AND (urodynamics)[All fields] (String 6) #29; ((spinal dysraphism)[All fields] OR (spina bifida)[All fields]) AND (functional assessment)[All fields] (String 7) #36; ((spinal dysraphism)[All fields] OR (spina bifida)[All fields]) AND (genetics)[All fields] (String 8) #51; The inclusion criteria were following article types: Meta-analysis, Randomized Controlled Trial, Review and Systematic Reviews.

Additional search in PubMed/MEDLINE was done for search terms (spina bifida)[All fields] and (ICF)[All fields] #10; (spina bifida)[All fields] and (multidisciplinary)[All fields] #84; (spina bifida)[All fields] and (guidelines)[All fields] #97 for all article types; and Methodology of "Physical and Rehabilitation Medicine practice, Evidence-Based Position Papers: the European position" produced by the UEMS-PRM Section #1.

Search in Grey literature (Supplementary material): #2 articles.

The recommendations draft and consensus followed the five-step Delphi procedure that was proposed by the Methodology paper.[11] Strength of Recommendations (SoR), and Strength of Evidence (SoE) grading was described as well in the Methodology paper.[11]

Recommendations were prepared according to what proposed in the Methodology paper[11] as defined by the professional practice committee of the UEMS-PRM section:

A. Generic recommendations;

B. Recommendations on PRM physicians' role in medical diagnosis using the ICD;

C. Recommendations on PRM physicians' role in diagnosis and assessment according to the ICF;

D. Recommendations on PRM management and process;

E. Recommendations on future research about best PRM professional practice in children and adults with spinal dysraphism.

Results

Systematic review

The electronic literature search identified 894 papers, from which 529 abstracts were selected and finally 50 articles were considered to produce this paper. A flow chart of the selection process is presented in Figure 1.

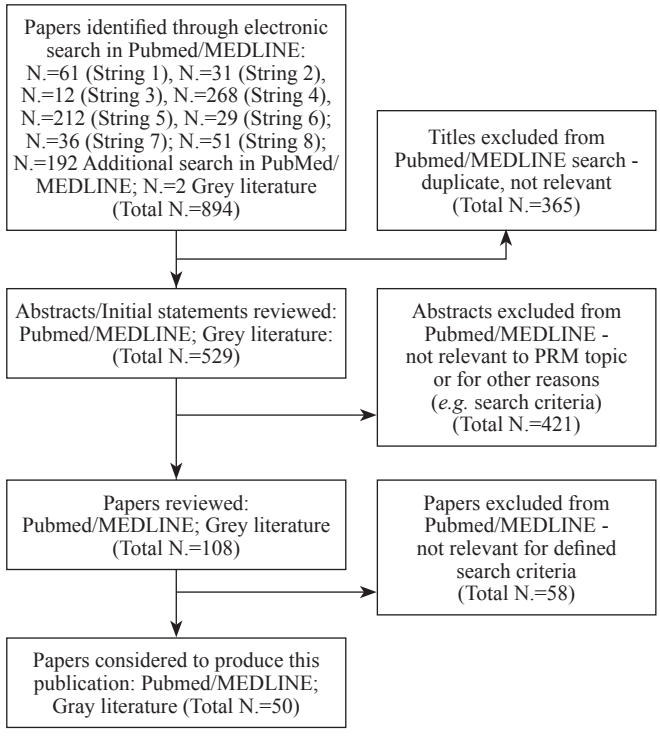

Figure 1.—Flow chart of the selection process.

Evidence synthesis

In this paper 15 existing guidelines were included that cover the broad aspects of the care for the people with SD. These guidelines can be considered on each level of care including primary, secondary and tertiary. Furthermore, they can be of great importance for caregivers and families.

The following issues emerged from the literature search:

• Maintaining functional capacity of musculoskeletal system with regards to the current neurological function.[7] With regards to the motor and sensory function that could be affected in the presence of neurological dysfunction, the physician should be able to predict the level of mobility that can be achieved.[10] This is of particular importance since mobility in pediatric population have impact on cognitive and psychological development.[10]

• Special attention should be given to the adjustment of health promotion and preventive interventions to the age of the persons with SD. Monitoring of caregiver burnout, child abuse or neglect, hearing and vision disturbances, pressure ulcers and skin cancer should be maintained., Patients' and caregivers' education on monitoring and prevention of secondary conditions such as pain of neurogenic and musculoskeletal origin, bowel and bladder dysfunctions, sleep apnea, etc. are also important.[12]

• The need of collaboration between families and clinicians in assessment of self-management readiness with a structured, planned, and incremental approach is recommended with a special emphasis on self-management and promotion of independence.[13] Family functioning should be directed towards parental and marital stress as well as maladaptation minimization when rising the child with SD, and to maximize family engagement in social activities and parental knowledge on SB and advocacy.[14]

• The neuropsychological assessment should be performed at pivotal moments of the individual's development in order to set strategies for learning as well as to improve outcomes for independent functioning.[15] The assessments performed in early childhood should include mobility, language and attention, as well as problem-solving skills and attitude of parents. The assessment in later childhood should address developmental needs focused on school adaptation and learning skills.[15]

• Males with SD are usually affected with sexual dysfunction. They can have increased frequency of bladder and bowel dysfunctions as well.[16] Females with SD usu-

ally could have specific needs and concerns in one or more issues such as: puberty, sexuality, pregnancy, childbirth, and menopause. Special attention in females thus should be focused on open communication, presence of decreased pelvic sensation, renal function assessment before conception, discussion of bowel care, management of seizures, mobility issues during pregnancy, preterm birth risks. Pulmonary function testing during pregnancy should be considered in females with kyphoscoliosis.[17]

Recommendations

The results of the Delphi procedure are listed in Table I and the overall view of the recommendations is presented in Table II.

A. Generic recommendations

1. The professional role of a PRM physician in the management of individuals with SD is to formulate, facilitate and govern an individually tailored rehabilitation program bearing in mind the age, type and level of the deformity, and the accompanying comorbidities and pathologies. The PRM physician needs to adopt the concept of integrated care in a multiprofessional and interdisciplinary managed team considering impairments, activity limitations and participation restrictions at all levels of individual and societal components.[3, 4, 13] [SoR:A; SoE:IV]

2. The rehabilitation interventions need to be implemented at all healthcare system levels (such as health care centers, hospitals, clinics, nursing homes, and home-care). The PRM physician's role throughout the rehabilitation program is to facilitate the involvement of the family and caregivers of the individual, particularly those with challenging circumstances (such as children and elderly, severe disability, cognitively impairments and those lacking capacity to consent for treatment). [SoR:A; SoE:IV]

B. Recommendations on PRM physicians' role in medical diagnosis using the ICD

3. The PRM physician should have specific and comprehensive knowledge of medical diagnosis and management of individuals with SD.[2, 6, 13, 18] PRM physician must be aware of the updated recommendations, guidelines, evidence-based interventions, and consensus statements in SD care. [SoR:A; SoE:IV]

4. The PRM physician needs to perform regular and timely follow-ups to observe and monitor functional progress and check for symptoms and signs of complications and comorbidities (such as pain, infections, psychosocial problems, cognitive difficulties, and problems related to the orthopedic, neurological, gastrointestinal, urological and reproductive system) both during childhood as well as adulthood.[4, 7, 16, 17] [SoR:A; SoE:IV]

5. The PRM physician needs to have specific expertise in performing and interpreting electrodiagnostic (electro-

TABLE I.—*Results of the consensus procedure.*

	Round	Number of recommendations	Accept	Accept with changes	Reject
1	1st vote	35	25.71%	62.86%	11.43%
	2nd vote	33*	54.55%	45.45%	0%
2		33	96.97%	0%	3.03%
3		32**	100%	0%	0%
4		32	100%	0%	0%
5		32	100%	0%	0%

*2 new added and 4 rejected after 1st vote of Round 1; **1 rejected after Round 2.

TABLE II.—*Overall view of the recommendations.*

Content	Number of recommendations	Strength of recommendations				Strength of evidence			
	N.	A	B	C	D	I	II	III	IV
Overall general recommendation	2	100%	0%	0%	0%	0%	0%	0%	100%
Recommendations on PRM physicians' role in Medical Diagnosis according to ICD	4	75%	25%	0%	0%	0%	0%	0%	100%
Recommendations on PRM physicians' role in PRM diagnosis and assessment according to ICF	2	100%	0%	0%	0%	0%	0%	50%	50%
Recommendations on PRM management and process	23	56.52%	39.13%	4.35%	0%	0%	17.39%	4.35%	78.26%
Recommendations on future research about best PRM professional practice in children and adults with spinal dysraphism	1	0%	100%	0%	0%	0%	0%	0%	100%
Total	32	62.5%	34.37%	3.13%	0%	0%	12.5%	6.25%	81.25%

myoneurography-EMNG) studies in patients with SD.[19] [SoR:B; SoE:IV]

6. The PRM physician needs to participate in transition care programs for adolescents with SD, and be involved in a closer follow-up for these individuals to promptly detect and manage the changes and other specific complications that could arise during this period.[4, 5] [SoR:A; SoE:IV]

C. Recommendations on PRM physicians' role in diagnosis and assessment according to the ICF

7. The PRM physician needs to perform a complete functional assessment including activities and participation with special attention to the presence of specific impairments (such as deformities of the musculoskeletal system, bladder and bowel dysfunction, hydrocephalus, and pain).[1, 2, 8, 20, 21] [SoR:A; SoE:III]

8. The PRM physician needs to use the World Health Organization (WHO) International Classification of Functioning, Disability, and Health (ICF) framework, the International Classification of Functioning, Disability, and Health for Children and Youth (ICF-CY), and/or (generic and/or specific) ICF Core Sets where appropriate for individuals with SD to collect and analyze the functioning components including activities (such as self-care, mobility, and communication) and participation (such as school, social, recreation, vocation and sports).[21-25] [SoR:A; SoE:IV]

D. Recommendations on PRM management and process

Inclusion criteria (when and why to prescribe rehabilitation interventions)

9. The PRM physician needs to prescribe and perform optimal patient centered rehabilitation program within multiprofessional team working in an interdisciplinary fashion for individuals with SD presenting with disability and participation restrictions.[3, 4, 9] [SoR:A; SoE:II]

10. The PRM physician needs to re-evaluate, modify and perform individualized rehabilitation treatment in individuals with SD when there is a progression of comorbidities, functional impairments and participation restrictions. Collaboration with other specialties and health care professionals is advised for making best interests decisions.[4, 9] [SoR:A; SoE:II]

Project definition (definition of the overall aims and strategy of rehabilitation interventions)

11. The PRM physician within the multiprofessional team needs to specify overall aims, design and procedures for rehabilitation interventions, monitor treatment courses and evaluate outcomes bearing in mind patients' age, and their needs and expectations. Furthermore, special consideration should be given to self-management and education of family members and/or caregivers.[26-28] [SoR:A; SoE:III]

12. The rehabilitation interventions should be implemented in every phase of the life cycle including transition for childhood to being adults. This is of importance due to the chance of getting lost in the system during the transition period. The individual's education level and capacity need to be taken in to account when discussing and planning interventions. Moreover, the use of e-health interventions for individuals with SD needs to be considered.[29, 30] [SoR:A; SoE:IV]

13. The PRM physician needs to follow established guidelines and practice recommendations, and participate in discussions with health professionals, patients, families, and caregivers while making evidence-informed decisions on health care interventions for individuals with SD.[31] [SoR:A; SoE:IV]

Teamwork (professionals involved and specific modalities of teamwork)

14. Rehabilitation of individuals with SD needs to be performed by a multiprofessional, interdisciplinary managed team. It can be provided in any setting or level of care (such as primary care, specialty care and community-based services).[32] [SoR:A; SoE:IV]

15. Depending on the individual needs and different settings, the multidisciplinary rehabilitation team should include the individual with SD, PRM physician, other relevant medical specialists (such as pediatricians, neurosurgeons, neurologists, orthopedic surgeons, urologists, internal medicine physicians, and primary care physician), rehabilitation professionals (such as physiotherapists, occupational therapists, nurses, neuropsychologists, speech and language therapists, orthotists, social workers, and community care workers), family members and caregivers.[15, 32-35] [SoR:A; SoE:IV]

PRM interventions

16. The PRM physician needs to have an active role in the management and prevention in early asymptomatic stages of complications and comorbidities in individuals with SD regardless of age. Assessing personal and environmental possibilities and/or barriers within multidisciplinary interventional programs could be of advantage in improving individual's participation in physical activity.[36] [SoR:B; SoE:IV]

17. The PRM physician needs to inform and provide counselling to the families about the importance of individual's proper care, possible medical and social needs. Moreover, the team of SD specialists and primary care providers should give consideration to individual's age-appropriate health promotion including adapted physical and recreational activities and participation.[12] [SoR:B; SoE:IV]

18. The PRM physician needs to participate in the multidisciplinary teams of pre- and postoperative care of individuals with SD who develop arthritis (such as end-stage degenerative process) aiming for pain reduction and function restoration.[37] [SoR:B; SoE:IV]

19. Incomplete bladder emptying in individuals with SD and neurogenic bladder can be managed by intermittent catheterization. Furthermore, individuals can be considered for intra-detrusor injections of botulinum toxin A (BTX-A) in the treatment of neurogenic detrusor overactivity that is refractory to antimuscarinics.[34, 38, 39] [SoR:B; SoE:IV]

20. The PRM physician needs to prescribe, perform and monitor effects of neuromodulation interventions by electrostimulation of the sacral nerve roots, tibial nerve, pudendal nerve, and dorsal genital nerves in individuals with SD and lower urinary tract dysfunction and/or neurogenic bowel.[34, 40] [SoR:C; SoE:IV]

21. Orthoses and mobility aids, such as walking aids and wheelchairs, are prescribed when needed for individuals with SD to improve biomechanical efficacy.[23] [SoR:A; SoE:IV]

22. Preventive measures including general preventive health, screening, recognition, management and adequate referral of common secondary or chronic conditions (such as hypertension, metabolic syndrome, and obesity); as well as skin care, and healthy life style activities[36, 41-43] should be promoted and incorporated in rehabilitation of individuals with SD. [SoR:B; SoE:IV]

Outcome criteria

23. The PRM physician needs to determine outcome criteria by performing physical examination of individuals with SD, and give opinion based on individuals' impairments, activity limitations and participation restrictions, bearing in mind age, associated comorbidities and complications. [SoR:A; SoE:IV]

24. The PRM physician and the rehabilitation team need to perform disability assessment of individuals with SD using different domains questionnaires including (but not limited to):[9]

a. Urogenital Distress Inventory (UDI6)

b. American Urological Association Symptom Index (AUA)

c. Incontinence Impact Questionnaire (IIQ7)

d. Wexner Faecal Incontinence Score (WFIS) [SoR:B; SoE:II]

25. The PRM physician with their team need to perform participation assessment of individuals with SD using questionnaires addressing psychosocial domains of functioning including (but not limited to):[9]

a. Depression Anxiety Stress Scale (DASS)

b. McGill Quality of Life Questionnaire (MQOL)

c. Brief COPE Scale (B-COPE)

d. Generalized Self-efficacy Scale (GSE) [SoR:B; SoE:II]

26. The PRM physician and their team need to perform quality of life and health-related quality of life assessments of individuals with SD with the following instruments including (but not limited to):[44]

a. Generic instruments:

- Pediatric Quality of Life Inventory (PedsQL)
- Child Health Questionnaire (CHQ Child)
- KIDSCREEN
- World Health Organization Quality of Life BREF (WHOQOL-BREF)
- McGill Quality of Life Questionnaire (MQOL)

b. Spina bifida-specific instruments:

- Health Related Quality of Life Spina Bifida (HRQOL-SB)
- Hydrocephalus Outcome Questionnaire (HOQ)
- Quality of Life Assessment in spina bifida for Children (QUALAS-C)
- Quality of Life Assessment in spina bifida for Teenagers (QUALAS-T)
- Quality of Life Assessment in spina bifida for Adult (QUALAS-A)
- Spina Bifida Pediatric Questionnaire (SBPQ)

c. Family Quality of Life instruments:

- Family Quality of Life (FQOL) [SoR:B; SoE:IV]

27. The PRM physician needs to use in clinical setting the Transition Readiness Assessment Questionnaire – Spina Bifida (TRAQ-SB) and other instruments for the care of individuals with SD. Additionally, such questionnaire can be of help in specific self-management skills acquisition in children and adolescents with SD.[45, 46] [SoR:B; SoE:IV]

Length/duration/intensity of treatment (overall practical PRM approach)

28. The PRM physician needs to follow recommendations, guidelines, consensus statements, and evidence-based po-

sition papers when deciding on the optimal length and duration of rehabilitation treatment and adequate intensity for individuals with SD keeping in mind different age groups and presence of comorbidities and pathological conditions. [SoR:A; SoE:IV]

*Discharge criteria (*e.g. *when and why to end PRM interventions)*

29. The PRM physician needs to consider discharge criteria based on an individualized patient centered approach bearing in mind individuals' overall and specific conditions in the acute and post-acute course of the rehabilitation program. Such approach is needed for both pediatric and adult populations. [SoR:A; SoE:IV]

Follow-up criteria and agenda

30. The PRM physician in the multidisciplinary setting needs to follow-up individuals with SD and facilitate provision of rehabilitation interventions and diagnostic modalities (such as neurophysiological, imaging techniques, urodynamics, and laboratory investigations) taking into consideration the patient's general health condition, age and the course of specific physical and cognitive challenges. This will enable timely recognition of early deterioration and subclinical or clinical changes.[27, 47-49] [SoR:A; SoE:IV]

31. The PRM physician needs to participate in and manage the regular follow-up of individuals with SD. This will enable inclusion of individuals with SD patients in rehabilitation programs taking into consideration the overall general condition, comorbidities, and physical, psychosocial and cognitive aspects of the condition. [SoR:A; SoE:IV]

E. Recommendations on future research about best PRM professional practice in children and adults with spinal dysraphism

32. Research areas of interest for the PRM physician needs to include but not limited to: diagnostic modalities, treatment options, assistive devices, home-care management including telerehabilitation, social inclusion and family participation.[7, 10, 14, 50] [SoR:B; SoE:IV]

Conclusions

The professional role of the PRM physicians requires specific expertise in the treatment of patients with SD to plan, lead and monitor the rehabilitation process in a multiprofessional setting through an interdisciplinary approach. Furthermore, as a complex anomaly, implementing new methods in early and timely diagnostics and preventing complications is needed in patients with SD. Also, implementation of the procedures and rehabilitation techniques with occupational therapy and orthoses as well as functional orthotic equipment is advised for functional recovery and better quality of life in all age groups. Furthermore, PRM physicians should participate in the assessment of the needs of these patients in the transitional phase from childhood to adulthood with special attention to the activity and the participation limitations. Therefore, the PRM physician's role is very important in a multiprofessional team because of the active role in treatment and follow-up across the entire life span. Moreover, the PRM physician should work on the implementation and, where existing, improvement of specialized rehabilitation services with the focus on promotion to the maximum possible functional level and integration in the community. This EBPP represents the official position of the UEMS PRM Section and describes the professional role of PRM physicians in children and adults with SD.

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Conflicts of interest.—The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

Authors' contributions.—Ivana Petronic Markovic and Dejan Nikolic contributed to database searching, data selection, analysis and interpretation; Carlotte Kiekens contributed to the study supervision; Ivana Petronic Markovic and Dejan Nikolic participated in manuscript drafting; Piotr Tederko, Minna Stahl, Stefano Negrini, Oksana Hdyrya, Mauro Zampolini and Carlotte Kiekens participated in data interpretation and manuscript critical revision; Dejan Nikolic submitted the paper. All authors read and approved the final version of the manuscript.

Acknowledgements.—The authors wish to acknowledge other members of the UEMS PRM Section Professional Practice Committee and other PRM delegates/experts involved in Delphi procedure for their valuable comments on this paper: A. Oral (Turkey), M.G. Ceravolo (Italy), P. Cantista (Portugal), E. Ilieva (Bulgaria), E. Varela Donoso (Spain), M. Lazovic (Serbia), D. Wever (Netherlands), J. Kujawa (Poland), M. Sivan (UK), A. Kucukdeveci (Turkey), I. Treger (Israel), F. Grubisic (Croatia), K. Hornacek (Slovakia), R. Crevenna (Austria), X. Hu (Sweden), J. Macfarlane (Ireland), F. Armakola (Greece), M. Leches (Luxembourg), N. Laktasic Zerjavic (Croatia), V. Golyk (Ukraine), P. Takac (Slovakia), A. Nulle (Latvia).

History.—Article first published online: May 16, 2022. - Manuscript accepted: May 2, 2022. - Manuscript revised: April 15, 2022. - Manuscript received: March 28, 2022.

Supplementary data.—For supplementary materials, please see the HTML version of this article at www.minervamedica.it