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William R. Black
Children's Mercy Kansas City

Lora L. Black

Jordan T. Jones
Children's Mercy Hospital

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Barriers to the Diagnosis, Care, and Management of Pediatric Patients With Ehlers-Danlos Syndrome in the United States: A Qualitative Analysis

William R. Black, PhD[1,2,3], Lora L. Black, PhD[1,3], and Jordan T. Jones, DO[4]

Abstract

Objectives: Ehlers-Danlos Syndromes (EDS) are a family of heritable connective tissue diseases. Primary practitioners are capable of diagnosing and managing EDS; however, few are knowledgeable and comfortable enough to see patients with EDS, resulting in delays in diagnosis and care. This study explores the barriers physicians experience with diagnosing, managing, and caring for patients with EDS, and potential resolutions to those barriers. *Methods*: As part of a larger online study, providers (n = 107) in the United States were asked to specify "What information would improve (their) comfort" in diagnosing, caring for, and managing EDS via open-ended questions. *Results*: Providers reported wanting clinical practice guidelines, in formats that were easily accessible and usable, information on their roles in the management of EDS, the best ways to coordinate with specialty care, and available specialty consultation. *Conclusions*: Providers overall are willing to diagnose and treat EDS; however, additional supports and training are needed.

Keywords

Ehlers-Danlos syndrome, adolescent, pediatrics, provider knowledge, diagnosis, treatment, qualitative

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Introduction

Ehlers-Danlos Syndromes (EDS) represent a family of heritable connective tissue disorders characterized by a number of physical features (eg, joint hypermobility, tissue fragility, and skin hyperextensibility)[1,2] and occurs in approximately 1 in 2500 to 5000 individuals worldwide annually.[3] There are currently 13 officially recognized distinct clinical subtypes,[2,4] with a 14th subtype that has been recently described but not yet officially classified.[5-7] Each EDS subtype is defined by major and minor criteria. Patients with EDS are prone to many co-morbidities across multiple body systems, including neurologic,[8,9] cardiovascular,[10] gastrointestinal,[11] dermatologic,[12] gynecologic,[9] and musculoskeletal issues.[13] Many with EDS suffer from chronic pain,[14] deficits with proprioception,[15] headaches,[16] anxiety, and depression.[16] The number and severity of co-morbid symptoms may lead to delays in diagnosis and challenges with diagnosis and management of EDS.[17] Delayed, missed, and incomplete diagnoses can greatly postpone family access to needed intervention and appropriate management strategies. Previously published research on barriers to EDS care suggests that diagnostic delays are at least partially attributable to several factors, including inadequate provider education and awareness of EDS, lack of known referral sources and specialists to whom suspected patients may be referred, and an increased provider burden to manage and provide care for patients with broad

[1]University of Kansas School of Medicine, Kansas City, KS, USA
[2]Center for Children's Healthy Lifestyles and Nutrition, Kansas City, MO, USA
[3]The Ohio State University, Columbus, OH, USA
[4]Children's Mercy Hospitals and Clinics, Kansas City, MO, USA

Corresponding Author:
William R. Black, Center for Biobehavioral Health, Abigail Wexner Research Institute at Nationwide Children's Hospital, The Ohio State University, 431 S. 18th St., Columbus, OH 43205, USA.
Email: William.Black@nationwidechildrens.org

and significant impairments.[18] Due to the breadth and intensity of these clinical concerns, intensive interventions may be needed for which primary practitioners may not feel adequately equipped to provide.[19] Further, primary practitioners have expressed a preference to refer patients to pediatric subspecialists to address individual system concerns and needs in the context of EDS rather than managing them within the primary care setting.[19] Additionally, there is a national shortage of pediatric subspecialists which may also affect access to accurate EDS diagnosticians and management.[20]

The absence pediatric subspecialists that are aware and comfortable with EDS necessitates other strategies to address diagnostic and management delays. There have been recent efforts to inform medical providers about EDS to aid in symptom management[21]; however, primary practitioners in our previous study reported a number of concerns around the diagnosis and management of EDS that affect their knowledge and comfort in managing EDS.[18] Further, these practitioners reported they are aware of EDS, but not familiar or comfortable with the 2017 International Diagnostic criteria for EDS. They also reported discomfort with developing care plans for patients with EDS, and are uncomfortable implementing care plans developed by a subspecialist.[18] As such, a lack of providers willing to diagnose and care for pediatric patients with EDS may be attributed largely to a lack of knowledge and training in the diagnosis and management of EDS.

Primary practitioners are positioned to provide care for chronic illness patients,[22,23] including EDS, by helping patients navigate the assessment process and initiate intervention, either in isolation or as part of larger health-care team that includes subspecialists. This is uniquely important to patients with EDS as many have high resource utilization across many healthcare systems and specialties that can lead to fractured care and unclear patient ownership,[24] Targeting primary and community practitioners may help patients and families access intervention earlier in their treatment, thereby reducing disease severity and potentially decreasing their reliance on[18] subspecialty care in the future. To address barriers to care, including a lack of knowledge and confidence around the care and management of EDS, EDS-specific educational materials and programs are needed. The objective of this study was to expand upon the findings from previous literature about provider barriers regarding EDS treatment[18] and to qualitatively investigate provider needs and preferences around EDS knowledge and education among medical providers in the United States, to inform the future development of physician educational programs.

Methods and Materials

Design

This study was conducted as part of a larger study,[18] in which REDCap electronic data capture tools hosted at Children's Mercy Hospitals were used to electronically collect information from providers in several areas, including their awareness of EDS, diagnostic practices of EDS, their level of comfort associated with the care, management, and education of EDS, as well as potential barriers prevented comfort with care, management, and education of EDS was administered to providers. The current study explores the qualitative responses while the results of the quantitative survey have been published previously.[18] The survey was electronically sent via email to a convenience sample of pediatric and family medicine practitioners and trainees at 2 tertiary care academic medical centers in the Midwest region of the United States. Inclusion criteria were currently being a provider of medical care in pediatrics or family medicine within the institution's city and the surrounding region, while exclusion criteria were not being a medical provider. Participants were asked to respond according to their personal experience and perspective. The survey was sent twice, in December 2021 and again in February 2022.

Providers were asked to provide free-written responses to several open-ended questions:

1) "What information would improve your comfort in diagnosing EDS?";
2) "What information would improve your comfort in caring for EDS?";
3) "What information would improve your comfort in managing EDS?"; "What information would improve education to families with EDS?";
4) "What topics would be helpful for an EDS workshop?".

Participants were able to skip questions, partially complete questions, complete the survey at a later time, or withdraw their consent by not submitting their answers.

Analysis

Responses were analyzed via thematic analysis to identify, analyze, and report themes found within the data[25] using Dedoose (Version 9.0).[26] Two team members (LXX [a PhD psychologist with 11 years of clinical work with chronic illness patients and 13 years of qualitative analysis experience] and WXX [a PhD psychologist with 15 years of clinical work with chronic illness

patients 4 years of qualitative analysis experience]) reviewed the qualitative responses and developed codes, which were discussed and agreed upon by team members. These team members then independently categorized responses into the codes. Any discrepancies in coding were discussed until consensus was reached. In the event consensus was not reached, a third team member (JXX [a male DO rheumatologist with 11 years of clinical work with chronic illness patients and 2 years of qualitative analysis experience]) reviewed the responses and codes to decide on final coding categorization. As there are no formal a priori power analyses for qualitative research no such calculations were conducted. In lieu of this, data saturation[27] was used as a marker for determining appropriate sample size.

After primary and secondary coding, themes were evaluated for similarity and uniqueness across the free-response prompts. It was noted that several primary themes emerged with consistency across the prompts, regardless of whether providers were being asked to comment on issues related to diagnosis, care, or management. Data saturation was met, as no new relevant data or themes emerged by the end of analyzing the qualitative responses.

Results

Participants

Of 107 survey responses analyzed, 73 (68%) provided at least one response to any of the 5 open-ended questions. The respondents were mostly pediatric residents (47%) followed by pediatricians (21%) and specialty physicians (11%). Most of the respondents (53%) had <5 years of experience while 25% had 10 or more years of experience.

Guides to Diagnostics and Practice

Several providers requested having access to "clinical practice guidelines" (CPGs) or standards of care, access to the "diagnostic criteria," and incorporating and hosting EDS CPGs to their hospital's intranet. Providers expressed interest in having a guide for what to do at various appointments:

> *"I feel like almost having a 'clinical practice guideline' such as what to look out for and ask about every visit would be great."*

Providers also expressed wanting guidelines for the diagnosis of patients with EDS, its care and care plans, and common management strategies and recommendations, including "common treatments used and how successful they are."

Providers also commented on the format and accessibility of the criteria, stating that it should be "easily accessed," brief, such as a "short" or "one sheet printout," a "Brief video or podcast to recap key diagnostic features and criteria," research, review, and journal articles, online resources such as UpToDate, and include "simple modules." Respondents also suggested that the diagnostic and care information be organized into clinical "algorithms" or a "flow chart or easy diagnostic tool."

Referrals and Involvement of Other Providers and Subspecialists

While respondents indicated areas of need that would improve their comfort and ability to diagnose, care for, and manage patients with EDS, several providers also spoke to the need to involve other providers. Many wanted information on "when and who to refer to" and expressed the need for more knowledge on how they fit within the patient's care plan, including "what needs to be communicated to specialist[s] and what I should be able to handle and how," "more knowledge of what my specific role in EDS care is as [Primary Care Provider]," and "what [Primary Care Provider]s should know and feel comfortable managing."

Similarly, providers reported wanting to have access to referrals and supportive specialists, such as "Physical therapist and support professionals with comfort and experience with condition" and other "specialists who should be involved with the child's care."

Experience and Exposure

Providers also expressed a need to have "more experience" or "more exposure" to working with patients with EDS:

> *"I would probably have to see this more often. . . I almost never diagnose someone with this (and they often come in with the diagnosis)."*

Providers want "Better familiarity with diagnostic criteria and subtypes of EDS," "repeating the diagnostic criteria multiple times to feel comfortable with it," while some felt they were able, but not comfortable in providing appropriate care to patients with EDS due to lack of exposure:

> *"I believe that I could identify the right resources that could guide me in my management. However, having limited to no exposure in managing patients with EDS, I am still not comfortable or confident in my skills."*

Consultation and Co-management With Specialists

Multiple providers expressed a desire to consult with subspecialists, specifically in regard to "guidance" and "collaboration" with rheumatology and genetics; and consultation with a specialist was noted to be important for both the initial diagnosis and ongoing management. The involvement of multiple subspecialists was important for barrier resolution for primary practitioners to care for and management patients with EDS, especially with development of care plans. Providers also noted an explicit desire for patient specific care plans, such as "an outline of what is the plan of care for the patient" and "communication or documentation from sub-specialist regarding plan of care for the patients" to improve their ability to care for and co-manage patients with EDS. For some providers, consultation with a subspecialist seemed almost mandatory to their comfort in managing patients with EDS:

"I feel comfortable managing these patient(s) with rheumatology consultant, but it is not something that I manage alone."

"We see kids with mild EDS for regular check up's etc. Managing the EDS specifically could be a challenge without the help of a specialist."

"Helpful to be able to reach out to specialists taking care of EDS pts with questions regarding Management of a patient with EDS."

Education

Similar to providers' desire to have short, accessible clinical guidelines, participants also requested handouts, review articles, and quick reference materials for broader self-education. This includes web-based resources for their own education, but also for information to be provided to families.

"Links to validated medical resources about this topic"

"Having educational material to give to patients following the encounter"

"Helpful websites for families"

Providers suggested formal means of acquiring more information on the diagnosis, care, and management of patients with EDS, such as through seminars, lectures, and grand rounds:

"Having it included as part of a formal didactic or during one of our various inpatient conferences would be great!"

"Any formal education or material specific to EDS."

Providers suggested modalities such as "small pearls lectures" and online "learning modules" to help address knowledge gaps in EDS care and management.

Workshop Topics for EDS

When asked to provide topics and areas of information that providers would like to see in EDS workshops, the aforementioned themes around diagnosis, care, and management were repeated. However, numerous providers plainly requested a workshop on the "diagnosis of EDS" and "diagnostic criteria," and clinical "tip-offs" for EDS, such as what to look out for, signs and symptoms, "when to suspect EDS," and common comorbidities and symptoms. Providers also requested workshops on how to treat and manage patients with EDS, both regarding existing management strategies and therapies, but also "protocols or recommendations," "monitoring protocols," and treatment guidelines to steer their practice. Respondents also expressed wanting to know what evaluation was needed as part of EDS diagnosis and care, including laboratory tests, imaging modalities, and screening procedures to be ordered by a primary practitioner. Other requested educational content is additional information on when patients should be referred to a subspecialist, how to connect patients with the correct subspecialists, and greater clarity on their role in conjunction with subspecialty or multi-specialty care.

Discussion

This post hoc, qualitative analysis of medical providers builds upon the themes observed in the previous manuscript,[18] but also provides more granular information about provider self-reported needs in the diagnosis, care, management, and education of pediatric patients with EDS. While many providers are aware of EDS, most are not comfortable with the diagnosis, care, or management of patients with EDS. Encouragingly, this study demonstrates provider willingness to improve their engagement with EDS patients, if they are provided with additional resources and supports. Here we show that many of the medical providers would like more clarity on diagnostic criteria, more exposure and experience with care and management of patients with EDS, guidance on consultation with subspecialists that are knowledgeable about EDS, and more educational options for knowledge attainment. Additionally, an EDS workshop that covers

many of these topics was also suggested by the providers surveyed. These requests from medical providers can now be used to shape the appropriate knowledge and the vehicle to deliver that knowledge, which will remove barriers and allow primary practitioners to diagnose, care for, and manage patients with EDS.

One of the most prevalent themes to emerge was that providers were uncomfortable with the diagnostic criteria for EDS. Many providers reported lack of knowledge of the 2017 International Classification Criteria for EDS, which discusses the diagnostic criteria for EDS. However, it has been proposed that children with joint hypermobility should not be assessed with the 2017 criteria until they have reached biologic maturity. A new diagnostic framework for pediatric joint hypermobility was recently published and recommends that pediatric patients should be assessed using a pediatric-specific framework versus the 2017 criteria if they have not yet reached biological maturity.[28] As this was published after the collection of our survey responses, we were unable to inquire about provider knowledge and understanding of the new pediatric framework; however, the addition of this new criteria for younger patients highlights the evolving and challenging nature of providing appropriate diagnosis and care for patients with EDS. Further, access to these resources does not immediately instill confidence and additional comfort is needed through exposure to patients with EDS. An option to gain exposure could be the implementation of shadowing or hands-on clinical care experience in an EDS clinic. While an opportunity like this could be difficult for busy primary practitioners, trainees could be required or given the opportunity to rotate through an EDS experience that includes educational materials, but also hands-on experience and the opportunity to help diagnose and manage patients with EDS. A similar option could be available for primary practitioners, but it could be modified to be less time intensive with a virtual option or focused workshop. The Ehlers-Danlos Society Extension for Community Health Care Outcomes (EDS ECHO) is a portfolio of teleconferencing programs developed around the principles and practices of Project ECHO® and has an aim to increase clinician awareness and ability to assess and manage patients with EDS.[29]

Other concerns raised pertained to subspecialty consultation and co-management of patients with EDS. Multidisciplinary care has been recommended in the management of patients with EDS due to the complexity and number of comorbidities that can be associated with EDS.[30] Additionally, there are multiple reports of how to development a multidisciplinary clinic and provide care for patients with EDS[31,32]; however, these options are not available in all areas, which may lead to diagnostic and management delays. Patients and families with EDS do report a willingness to travel to obtain subspecialty or EDS-specific care,[24] but this comes at a significant time and cost-burden to families and is not a sustainable solution to improve consistent and available care and management of patients with EDS. Clear guidance needs to be established on co-management of patients and symptoms, and consultation with subspecialists that can be applied to any community regardless of resources available. The delineation of care and management that can be applied to each situation may then allow patient and provider to better understand ownership of specific issues and where general care is obtained as this is not clear to many patients and providers.

Universal, easily accessible education is needed to support the providers that do care and manage patients with EDS, but also to help those that need extra support in their care and management. There were multiple requests for a workshop that could delve into diagnosis and management of patients with EDS, but also many requests for clinical guidelines that could be easily accessible to the practitioner. Unfortunately, at the current time there is a strong need for more evidence-based approach to therapy[33] and need for consensus guidelines for care and management. Additionally, the needs of each community are likely different, but EDS-specific workshops could be developed that walk primary practitioners through diagnosis, care, and management of patients with EDS. These workshops could be offered routinely or even via virtual option (such as EDS ECHO) to increase opportunities to participate. Workshops could also bring providers with similar interests together to develop multidisciplinary care or help develop referral lines for improvement in co-management and better understanding of consultation for patient need.

This study has several limitations which may limit the generalizability of results. Namely, this study was conducted locally and results may not be applicable to practitioners in different geographical regions and to healthcare systems outside of the United States. Further, this study was conducted as an institution which has a multidisciplinary EDS clinic that serves the region; the availability of this resources may bias providers in both increased awareness and potentially a greater reliance on subspecialty care as a referral source.[31] We expect the themes reported in this study to be present and potentially more profound in areas without a multidisciplinary EDS clinic. Also, this sample includes a large number of trainees, which are earlier in their career, and they may be more interested to participate in EDS care compared to established clinicians that may have less availability or interest. However, primary practitioners have seen a growing role in the ongoing care and management of

pediatric chronic disease.[34] Finally, we did not use validated surveys in the current study which may limit the validity of how the information was assessed. However, no validated surveys currently exist that assess provider knowledge about EDS thus the questions were created by the researchers based on clinical expertise.

Conclusion

The themes observed in this study provide more granular information about provider needs in the diagnosis, care, management, and education of pediatric patients with EDS. We show that medical providers want clarity on the diagnostic criteria, more experience with patients with EDS, guidance on consultation with subspecialists, and educational options for knowledge attainment. The results of this study also show that providers are willing to participate in EDS care with the requisite knowledge, available tools, reference information, and management supports to do so. The themes observed may provide a framework to develop an EDS workshop or multiple educational modules that covers many of these topics, which could lead to fewer barriers and more comfort in diagnosis, care, and management of pediatric patients with EDS. The pursuit of such educational initiatives may increase provider engagement in EDS care and management immediately with additional resources and supports provided to practicing clinicians, but also long-term with providing resources to current trainees and early-career practitioners.

Author Contributions

WRB: contributed to design; contributed to acquisition, analysis, and interpretation; drafted manuscript; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring itegrity and accuracy. LLB: contributed to conception and design; contributed to analysis and interpretation; drafted manuscript; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring itegrity and accuracy. JTJ: contributed to conception and design; contributed to acquisition, analysis, and interpretation; drafted manuscript; critically revised manuscript; gave final approval; agrees to be accountable for all aspects of work ensuring itegrity and accuracy.

Declaration of Conflicting Interests

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Ethical Approval and Informed Consent

This work was conducted in accordance with the Declaration of Helsinki. Institutional review board approval was obtained from Children's Mercy Kansas City IRB (IRB Study ID: 16060435). All respondents consented when they voluntarily completed the survey and no verbal or written consent were obtained, in accordance with the minimal risk nature of the study, and with approval from the IRB. Data were de-identified prior to publication and no personal or identifiable information was deemed to be in the final manuscript by the authors. Thus, no written consent for publication was obtained from participants.

ORCID iDs

William R. Black https://orcid.org/0000-0002-1060-0187

Jordan T. Jones https://orcid.org/0000-0003-0986-4991

Supplemental Material

Supplemental material for this article is available online.

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