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REVIEW

Psychosocial considerations in pediatric autoimmune liver disease

Brianna C. M. Wellen[1,2] | **Henry C. Lin**[3] | **Jacklyn E. Stellway**[1]

[1]Division of Pediatric Psychology, Doernbecher Children's Hospital, Department of Pediatrics, Oregon Health & Science University, Oregon, USA

[2]Department of Psychology, University of Utah, Salt Lake City, Utah, USA

[3]Division of Gastroenterology, Department of Pediatrics, Oregon Health & Science University, Oregon, USA

Correspondence
Jacklyn E. Stellway, Division of Psychology, Pediatrics Department, Oregon Health & Science University, Portland, OR, USA.
Email: stellway@ohsu.edu

BACKGROUND

Pediatric autoimmune liver disease, including autoimmune hepatitis (AIH) and autoimmune sclerosing cholangitis (ASC), is a group of chronic liver conditions requiring life-long management.[1] The medical management involves long-term use of corticosteroids with or without immunosuppressive therapy, both of which carry risk of side effects. Some children do not respond to standard therapy and liver disease may progress, ultimately requiring a liver transplant. Continued medication adherence and clinical follow-up are two essential factors in long-term disease management.

PSYCHOSOCIAL IMPACT

Several studies have demonstrated that youth with autoimmune liver disease report lower health-related quality of life (HRQOL) than healthy controls.[2] Higher levels of mental health concerns are associated with lower treatment adherence in these youth, although the causal direction of this relationship is unclear.[3] Although research is sparse in pediatrics, rates of depression and anxiety are known to be higher in adults with AIH compared to the general population.[4]

Similar to other chronic illnesses, emotional distress and impairment associated with adjustment to AIH is likely related to a variety of factors, including the necessity for frequent medical visits and medication management challenges.[5] Additional psychosocial considerations for pediatric AIH include the uncertainty and fear around prognosis or disease progression (e.g., potential for necessity of liver transplant later in life), as well as unpleasant side effects of corticosteroids known to affect HRQOL[6] and which may affect adherence. In studies of diseases with similar disease-related uncertainty, such as inflammatory bowel disease (IBD), higher rated levels of uncertainty are related to poorer adjustment to illness[7] and increase in depressive symptoms.[8]

Given the chronic nature of AIH, the potential for disease progression, and necessity for long-term medication use and consistent medical follow-up, psychosocial considerations are an important but underappreciated aspect of care.

STRATEGIES FOR ADDRESSING WELLBEING AND ADHERENCE

To mitigate the effect of AIH on quality of life and promote treatment adherence, medical professionals can consider the following recommendations. Strategies should be considered within two areas: proactively addressing psychosocial concerns and interventions for identified concerns.

Proactive considerations

Managing uncertainty

Addressing ambiguity regarding disease progression (e.g., future necessity of liver transplant) can occur through proactive discussions with children and caregivers. Even when gastroenterologists cannot provide definitive information regarding prognosis, it is helpful to allocate time to listen and validate patient concerns. Additional strategies include encouraging families to focus on aspects of disease management that are within their control by developing specific and attainable behavioral goals (e.g., daily medication reminders and attending follow-up appointments). By emphasizing behavioral successes, providers can support patients and families in feeling accomplished with their participation in disease management, thereby reducing distress and impairment.[9]

Psychoeducation

Gastroenterologists/hepatologists are encouraged to discuss psychological and functional aspects of AIH at the time of diagnosis to mitigate its impact. Psychoeducation should involve explicitly outlining activities in which the patient can still participate (and encouraging this participation; e.g., school, sports, travel), problem-solving barriers to participation, and identifying systems of social support and engagement with the AIH community. Adherence to medications, participation in a child's usual activities, and cultivating a strong system of social support can be protective against adverse psychosocial impacts of chronic illness.[10] Additionally, adherence to medications may be negatively impacted by distressing side effects of corticosteroids. A discussion of potential side effects, and plan for mitigation, should occur between provider and patient.

During initial psychoeducation discussions, it is important for medical providers to create a safe and comfortable environment for families to ask questions and express concerns. This conversation can be fostered by withholding judgment and answering questions earnestly and thoughtfully. A supportive discussion promotes patient trust in the medical team and improves patients' recall of important treatment information.[11]

Assessment

Proactive strategies can also include assessment of risk factors that may contribute to poor disease adjustment. Youth demonstrating more pre-morbid challenges (e.g., behavioral or emotional concerns, limited access to resources) may have more difficulty adjusting to the chronic illness diagnosis and management.[12] The assessment should include questions about pre-morbid behavioral and emotional functioning (of both parent and youth), logistical/socioeconomic barriers (e.g., health insurance, transportation), health literacy, family functioning, and social support. Assessment tools, such as the PedsQL Medicines Scale,[13] are available. Additionally, the child's developmental stage should be considered when evaluating and intervening on psychosocial factors in AIH (e.g., Reference[14]). For younger children, assessing parental functioning may be a priority, whereas for older adolescents, evaluating and intervening at the level of the adolescent patient should be prioritized. This assessment can help providers to direct patients and their families to appropriate supports (Table 1).

Health-care transition (HCT)

Assessment of health-care self-management and provision of corresponding interventions is also warranted for AIH patients nearing adulthood. HCT is the process of assessing, educating, and supporting young people as they prepare to move from child- to adult-centered health-care. Studies have found that higher rates of medical non-adherence and increased morbidity and mortality occur immediately following the transition to adult care in chronic disease populations similar to AIH (e.g., IBD;[15]). Poor adherence, poor disease management, and limited disease knowledge have been noted by adult hepatologists as the greatest barriers to optimal health-care transition, and these challenges can be related to poor/limited preparation for transition.[16] To improve the transition period, experts recommend implementing a systematic transition policy, frequent assessment of adolescents' transition readiness and barriers (e.g., knowledge, financial), and a planned process for education and counseling of adolescents and young adults nearing transfer to adult gastroenterology.[16] Recommendations for assessment and transition planning are summarized in Table 2.

Interventions for identified concerns

Adherence challenges

Addressing difficulties with adherence to medication regimens is not a one-size-fits-all approach. These challenges may vary depending on family factors, including patient and caregivers' knowledge and understanding of the disease, psychological barriers (e.g., mental health concerns, stigma, uncertainty), and logistical barriers.[11] Individualization of interventions is recommended based on an assessment of relevant adherence barriers.[19] It is common for multiple barriers to interact, leading to a necessity for more complex interventions. For example, the uncertainty regarding medication efficacy and disease progression may lead to low prioritization of treatment regimens (psychological and medical barriers). This challenge may be further complicated for a family with limited resources who may allocate finances to other critical necessities

TABLE 1 Psychosocial challenges/barriers and recommended strategies

Challenge	Potential impact on AIH management	Strategies for medical teams	Referral
Logistical and Socioeconomic • Stable housing and meals • Adequate health insurance • Transportation • Child care	Adherence Challenges: limited resources and necessity for prioritization of basic needs (e.g., having housing), attending visits may not be logistically feasible (e.g., time of work) Impact on Wellbeing: added stress to existing challenges with basic needs	• Increase flexibility (offer a variety of dates/times for appointments, choice of virtual or in person) • Find local providers if possible	Social work: Help to find resources Disability services: Connect with Medicaid, other statewide supports, school accommodations
Health Literacy and Knowledge • Communicating with doctors • Managing health insurance • Understanding disease • Understanding management recommendations	Adherence Challenges: difficulty problem-solving and advocating, not taking medication when feeling well, avoidance of health-care visits, lack of understanding of necessity of seeking health-care Impact on Wellbeing: more uncertainty and related distress	• Give clear, simple, straightforward directions for attaining medication, taking medication, and when to follow up with doctor • Give information in multiple formats • Allocate additional time for teaching about illness	Social work: teach health literacy, assist in health-care management Disability advocate: assist in health-care management
Mental Health • Mood concerns (anxiety, depression) • organization and memory (ADHD, developmental delays, intellectual disability) • Parent mood concerns	Adherence Challenges: lack of motivation or organization, limited reminders from parents Impact on Wellbeing: More intense emotional concerns surrounding diagnosis, prognosis, and disease progression	• Suggest and help to problem-solve strategies for improving health behaviors, such as scheduled reminders, calendars, pill boxes • Take time to answer questions related to worries and concerns and provide validation	Behavior therapist: teach behavioral techniques to increase healthy behaviors, problem-solving, address mood concerns Counselor: teach to coping skills for chronic illness PCP or psychiatrist: prescribe psychiatric medications
Social Support • General family functioning • Parent/child relationship • Social/peer relationships • Caregiver support system	Adherence Challenges: family conflict increasing child oppositional behavior, lack of organization, limited help with management behaviors Impact on Wellbeing: Caregivers or child may become more overwhelmed with managing chronic illness, feel alone in disease management	• Work with parents and child to problem-solve challenges around medication adherence (e.g., who sets reminder, parents' involvement) • Validate concerns	Parent and child support groups: increase social support Family therapist: address family conflict and functioning

(a logistical barrier). If multiple or interacting barriers are identified, connecting the family with social work or behavioral health/psychology may be warranted (Table 1).

Mental health and emotional distress

Patients and families may endorse or display difficulty coping with AIH at follow-up visits. Although emotional distress related to a chronic illness is common and at times expected, an assessment of these concerns is crucial to identify patients requiring additional support. Given that patients and families may be hesitant to start a conversation about mental health concerns with their medical provider, physicians can begin by conducting a brief assessment. This assessment may include questions about mood, fears, daily functioning (e.g., school, friends, and hobbies), sleep, and coping strategies. For

TABLE 2 Health-care transition (HCT) guidelines

Key area for transition readiness	Disease and medication knowledge	Autonomy/independence in health-care management	Health and lifestyle
Health-care engagement by age—individualized consideration should be made for developmentally-appropriate expectations based on cognitive and social–emotional functioning			
Early adolescence (12–14)	• Learning to describe specifics of their liver disease • Learning about the importance of medical adherence • Can name medications, dosing, schedule & side effects	• Plan for one-on-one time with their provider • Can answer at least 1 question during health care visit	• Learning healthy lifestyle choices (e.g., food, exercise, sleep, peer influence) • Can articulate their needs in school and other environments (e.g., sports)
Mid adolescence (14–17)	• Demonstrates knowledge of medical history and answers disease-related questions during visits • Developing independent medication management (i.e., dosing, schedule) • Know names and purposes of necessary labs and tests • Understands the risk of medical non-adherence • Learning about filling prescriptions	• Increasing one-on-one time with provider during visits • Scheduling medical visits, communicating with medical team (e.g., phone calls, EMR messages) • Knows names and roles of medical providers; knows how to contact medical team • Learning about insurance and medical bills	• Understands the impact of drugs and alcohol on liver condition • Advocates for their needs and accommodations in school and other activities (e.g., school, sports) • Discussing fertility/ sexuality with medical providers
Late adolescence (17+)	• Knows their medical history and answers disease-related questions • Independent medication management (i.e., dosing, schedule) • Can identify contraindicated medications • Arranges and follows through with necessary labs and tests • Arranges for prescriptions and refills • Receives medical summary and makes arrangements for an adult gastroenterologist	• Independent clinic visits with provider (without parents) • Demonstrates understanding of insurance coverage and paying medical bills; who to contact with questions • Communicating with medical team; scheduling visits	• Planning for after high school (i.e., college, work) and knows how to access resources (e.g., FMLA) and accommodations (e.g., college disability services) • Discusses with provider lifestyle choices, impact of drugs and alcohol on liver condition
Role of Medical Provider/Team	• Ongoing assessment of transition readiness and • Focus attention on the patient rather than the guardian when providing any explanations • Engage youth and parents/ caregivers in setting specific goals (e.g., filling prescriptions, getting lab work, contacting medical team); document and check on progress at next visit	• Encouraging independence, speak to child directly; increasing interaction between teen and health-care provider • Discuss importance of preparing the patient for health-care independence and address any anxiety • Discuss plans for insurance coverage • Encourage/facilitate scheduling visit with adult gastroenterologist	• Provide information about diet, fitness, drugs, alcohol, sexuality/ fertility • Inform patient of legal guidelines around information that can/will be shared with parent/guardian, and patient's rights to health care decisions after age 18 • Provide patient with medical summary for work, school, and HCT to adult gastroenterologist
Clinic HCT Guidelines	Six Core Elements to establish a HCT Program (GotTransition.org): 1. Create a policy/guide 2. Track and monitor progress 3. Assess self-care skills (transition readiness); use a standardized transition readiness assessment (TRA) 4. Develop a HCT plan with patient and family; transition planning 5. Transfer to an adult gastroenterologist 6. Confirm transfer and assess patient/family feedback		

(Continues)

TABLE 2 (Continued)

Key area for transition readiness	Disease and medication knowledge	Autonomy/independence in health-care management	Health and lifestyle
Transition readiness assessments	Transition Readiness Assessment Questionnaire (TRAQ;[17])—A 29-item patient-report measure with two domains: self-management and self-advocacy STARx questionnaire[18]—An 18-item patient- and parent- report measure with three subdomains: communication with medical provider, disease knowledge, and self-management Transition Readiness Assessment from Got Transition		

Note: Information in this table was adapted from Got Transition, NASPGHN Healthcare Provider Transitioning Checklist, and Improve Care Now.

youth and families with expected levels of emotional distress, feeling heard, validated, and supported by their gastroenterologist is often sufficient.[11] If a medical provider is concerned about more acute emotional distress or disruption to daily functioning, they are encouraged to consult with a mental health provider, preferably a professional with training in pediatric chronic health conditions. If a child is experiencing severe functional impairment (e.g., anxiety, avoidance of activities, symptoms of depression or post-traumatic stress) referral to a mental health provider is recommended.

SUMMARY

The management of pediatric autoimmune liver disease, similar to other pediatric chronic illnesses, is impacted by psychological and social factors of youth and their families. Medical teams can mitigate psychosocial distress and barriers to medical adherence through brief assessments and supportive care strategies.

CONFLICT OF INTEREST

Nothing to report.

ORCID

Brianna C. M. Wellen https://orcid.org/0000-0002-9371-690X
Henry C. Lin https://orcid.org/0000-0002-3927-6835
Jacklyn E. Stellway https://orcid.org/0000-0002-4078-7050

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