



HHS Public Access

Author manuscript

Am J Ophthalmol. Author manuscript; available in PMC 2019 October 11.

Published in final edited form as:

Am J Ophthalmol. 2018 July ; 191: xvi–xxiv. doi:10.1016/j.ajo.2018.03.028.

CHRONIC ANTERIOR UVEITIS IN CHILDREN: PSYCHOSOCIAL CHALLENGES FOR PATIENTS AND THEIR FAMILIES

Dr. Delana M. Parker, Dr. Sheila T. Angeles-Han, Dr. Annette L. Stanton, Dr. Gary N. Holland

Department of Psychology, University of California, Los Angeles (UCLA), Los Angeles, CA (Drs. Parker, Stanton), the Cincinnati Children's Hospital Medical Center and Department of Pediatrics, Division of Rheumatology, University of Cincinnati, Cincinnati, OH (Dr. Angeles-Han), the Ocular Inflammatory Disease Center, UCLA Stein Eye Institute (Dr. Holland) and the Departments of Ophthalmology (Dr. Holland) and Psychiatry and Biobehavioral Sciences (Dr. Stanton), David Geffen School of Medicine at UCLA, Los Angeles, CA

Abstract

Purpose—To describe issues of concern to children with chronic anterior uveitis; to consider the psychological impact of chronic anterior uveitis on children's lives; and to understand the effect of a child's chronic illness on other family members.

Design—Expert commentary.

Methods—Author experiences were supplemented by a review of pertinent medical literature and by consideration of content from semi-structured, separate patient and parent interviews.

Results—Vision loss and the fear of blindness are not the only stressors for children with chronic anterior uveitis and their families; of additional concern are the burdens of examinations and treatment regimens, as well as drug toxicities. Children with chronic anterior uveitis experience medical, academic, interpersonal, psychological, and developmental challenges. The impact of disease extends to other members of a patient's family as well; parents experience challenges in similar domains. Problems with adherence to medical regimens are common. Both the disease and its treatment affect quality of life, and can interfere with successful management of disease and transition to autonomy in adulthood, as reported for other chronic conditions. Coping processes vary greatly between different families.

Conclusions—Eye examinations and the rigors of long-term treatment often influence the psychosocial health of patients and families; physicians who are aware of these issues can help patients and families cope with chronic illness and may improve outcomes. Further psychosocial

Address for Correspondence: Gary N. Holland, M.D.; UCLA Stein Eye Institute; 100 Stein Plaza, Los Angeles, CA 90095-7000. uveitis@jsei.ucla.edu.

Dr. Parker is now associated with Department of Anesthesiology, Perioperative, and Pain Medicine, Stanford University School of Medicine, and Stanford Children's Health, Palo Alto, CA.

Financial Disclosure: The authors have no financial disclosures. Copyright for the Effect of Youngsters' Eyesight on Quality of Life (EYE-Q) questionnaire is held by Emory University, Atlanta, GA; Dr. Angeles-Han has received no royalties related to use of the EYE-Q questionnaire.

Publisher's Disclaimer: This is a PDF file of an unedited manuscript that has been accepted for publication. As a service to our customers we are providing this early version of the manuscript. The manuscript will undergo copyediting, typesetting, and review of the resulting proof before it is published in its final citable form. Please note that during the production process errors may be discovered which could affect the content, and all legal disclaimers that apply to the journal pertain.

research to understand the experiences of children dealing with chronic anterior uveitis is warranted; this commentary can serve as a foundation for development of age- and disease-specific research questions.

Chronic anterior uveitis, the most common form of uveitis in children, presents complex management problems for pediatric ophthalmologists, uveitis specialists, and pediatric rheumatologists.^{1–3} As many as two-thirds of children with chronic anterior uveitis have associated juvenile idiopathic arthritis (JIA), although it can occur with other systemic diseases, and it can occur in isolation.¹ Vision-threatening ocular complications of chronic anterior uveitis are particularly common in children; they include band keratopathy, synechiae and inflammatory membranes, secondary cataracts, uveitic glaucoma, hypotony, and macular edema.⁴ In addition, uveitis in children is associated with unique risks, such as growth retardation, if treatment involves systemic corticosteroids, and amblyopia, if there is vision deprivation from complications. Many, if not most children with chronic anterior uveitis require systemic immunomodulatory therapy to maintain control of inflammation. With current medical therapies and surgical management of complications, 80–90% of children with uveitis retain good visual acuity.⁵ While most children with chronic anterior uveitis will avoid severe vision loss, disease management itself often exacts a toll on the quality of life and functioning of patients and their families, who must manage the emotional sequelae of living with a burdensome disease.

In their relatively brief encounters with children who have chronic anterior uveitis, ophthalmologists naturally focus on disease findings and treatment recommendations. As with many chronic illnesses, though, disease status does not adequately convey the extent to which uveitis influences the lives of patients and their families. Astute ophthalmologists may recognize that disease findings and vision are not the only factors that matter to families, and can even become secondary to more immediate concerns, such as the expense and complexity of treatments and the disruptions associated with frequent examinations. Other issues, including psychological adjustment, adherence to medical regimens, and family interactions may also dramatically influence a patient's course of disease. As such, these issues should become a clinical focus for physicians as well. Such psychosocial considerations have been studied in the context of other chronic illnesses, but only a few have focused on children with uveitis.^{6–9}

Our goal in preparing this Perspective was to draw attention to the important and often overlooked psychosocial demands encountered by children with chronic anterior uveitis and their families. We also sought to characterize the many features of the disease and its treatment that result in these challenges. Commentary is based primarily on the experiences of a pediatric rheumatologist (STA-H) and a uveitis specialist (GNH), both of whom have many years of experience in caring for children with uveitis. The psychosocial implications of these observations are interpreted with the assistance of two health psychology researchers (DMP, ALS). As an additional measure of validation, the authors drew upon the results of interviews with children who have chronic anterior uveitis and their parents, as described below. Selected information and quotes from those interviews are used to illustrate the concepts being presented. We further supplement our observations with data from a review of the literature on quality of life among people, including children, with chronic

diseases. Situating our observations in the context of broader pediatric psychological research allows recommendations for future research and management of psychosocial concerns during clinical interactions.

Exploratory Interviews

We supplemented our clinical observations with a set of exploratory, semi-structured interviews of children with chronic anterior uveitis from the practice of one of us (GNH) and their parents. Two of us (DMP, ALS) conducted all interviews without input from the patients' physicians. DMP, at the time a pre-doctoral graduate student in clinical psychology, conducted all child interviews; ALS, a professor in clinical and health psychology, conducted all parent interviews. Interviews were determined to be exempt from review by the UCLA institutional review board.

Twelve parents (6 mothers, 6 fathers) and 6 children with chronic anterior uveitis (4 girls, 2 boys; current ages 4–21 years; 4 non-Hispanic white, 1 African-American, 1 Hispanic) from 10 different families were invited to be interviewed; none declined to participate. Each interview was conducted separately in a private room at the UCLA Stein Eye Institute or by telephone. Interviews were recorded and the qualitative information extracted from the interviews was interpreted in the context of existing psychosocial research on pediatric chronic illness. Descriptions of issues in the psychosocial adjustment of patients and parents were organized under several life domains.

CHILDREN'S EXPERIENCE OF UVEITIS

Throughout our experiences with children who have chronic anterior uveitis, we have observed that many experience medical, academic, interpersonal, psychological, or developmental challenges that affect their well-being (Table 1). In general, these observations correspond to reports in the medical literature relevant to children and to chronic disease states. DeCarlo and associates reviewed comments from focus groups of children with vision loss caused by ocular conditions other than uveitis (and their parents) and found that comments from 510 (44%) of 1163 children and 1078 (55%) of 1952 parents dealt with the following content categories related to quality of life: psychosocial; school-related; expectations and frustrations; dependency; and participation.¹⁰ Sen and associates conducted semi-structured interviews of 10 children with chronic uveitis who were 6–18 years of age, and identified comments across six domains: impact of treatment; ocular complications; impact on school; impact outside of school; social factors; and emotional reactions.¹¹ The sections that follow provide a summary of the demands faced by children and parents, based on our composite clinical impressions, with supporting evidence from recorded interviews and information in the medical and psychology literature.

Medical Challenges

Children with chronic anterior uveitis face various challenges related to the disease and its complications, and to its medical and surgical management. Patients may be bothered by floaters, and despite aggressive treatment, some patients continue to experience transient or permanent vision loss.^{1,5} Miserocchi and associates report that uveitis is associated with

worse physical and mental health-related quality of life in Italian adults, especially among women, older individuals, and those with low visual acuity (worse than 20/40) or longer disease duration (>6 years).¹² The same appears to be true among children. Angeles-Han and associates demonstrated that visual impairment among children with chronic anterior uveitis is related to worse overall quality of life, as measured by the Pediatric Quality of Life Inventory (PedsQL), version 4.0.^{6,13–15}

Additional important medical stressors arise in this population. Management of uveitis can be demanding, often involving complicated examinations, which can be frightening for young children, and complex regimens of topical medications and systemic immunomodulators, which can be difficult to follow. Patients may need multiple eye drops (corticosteroids, glaucoma medications, dilating agents), sometimes with frequent instillation and different regimens for each eye. Regimens can change often. Parents commonly say that their children resist eye drop administration, out of fear or because the medications sting. Young children, who find patches to be embarrassing or annoying, may resist treatment of amblyopia, which is sometimes necessary because of uveitic complications that result in visual deprivation in one eye.

Many of the long-term systemic medications used to treat chronic anterior uveitis, including methotrexate, infliximab, and adalimumab, require intravenous infusion or subcutaneous injection, the latter usually administered at home by the parent. The prospect of a “shot” is a well-known stressor for children. Furthermore, possible drug toxicities require monitoring through regular physician visits and laboratory tests. Ophthalmologists may not fully appreciate the impact of these factors on a patient’s well-being. A cursory question about how a child is tolerating weekly methotrexate injections may be met with what the child feels is an obligatory “OK”, when in fact the medication is causing substantial gastrointestinal upset, malaise, and fatigue. It is worth probing more deeply, if medication intolerance is suspected. Parents may also be reluctant to question the use of a particular medication, feeling that its continuation is essential, when in fact better tolerated and equally effective alternatives may exist.

Parents generally report that children, especially the very young, have difficulty understanding treatment regimens. Because adherence to these regimens is crucial for long-term control of inflammation and prevention of complications, most parents appropriately assume primary responsibility for administration of medications and monitoring of treatment, but some give even very young children limited responsibilities, such as holding the eyes open “like a big girl.” Children often reported involvement in the administration of their own eye drops beginning in middle school, although some parental monitoring usually continues through high school. In contrast, we occasionally find that parents have delegated eye drop administration to very young children, without supervision or monitoring, raising concerns about compliance and correct instillation.

Academic Competence

Some children report vision-related problems that affect functioning at school, including difficulty seeing the board in class (especially when teachers write in lighter colors or when lettering is not bold and precise); reading books with small print; and completing

schoolwork independently. In the current era of aggressive medical therapy, these difficulties do not appear to be pervasive, but unfortunately, when they do occur, not every student notifies teachers of the problems.

Children with uveitis may need to miss numerous complete or partial school days for eye examinations or treatments. Children may miss school for extended periods because of eye surgery related to complications. Many children describe difficulty compensating for missed course work, and these difficulties generally increase as children face more rigorous academic demands when they get older. School absence is a problem with other pediatric diseases as well,¹⁶ but visual impairment can compound these problems for those with uveitis. Unknown is whether disruption of a student's schedule by visits to the school nurse's office for eye drops or other medications also has an impact on academic performance.

Interpersonal Relationships and Recreation

We have observed that uveitis interferes with children's normal social and leisure activities to varying degrees. Parents report that changes in vision and the demands of treatment can strongly influence children's daily lives, affecting the ability to take part in play, and later, the ability to drive. Families describe periods of disruption after methotrexate injections, because of fatigue or nausea and vomiting. Although participation in some sports can be contraindicated because of disease or treatment, children and parents report numerous options for physical activity through other sports or recreational pursuits. Reflecting a sentiment commonly endorsed by parents, one mother promoted the idea that having uveitis is "not an excuse not to live your life well."

Disease-related absences from school result in missed opportunities to socialize with peers. Some children must avoid certain activities, such as swimming after eye surgery, or even sleepovers with friends, if it conflicts with treatment regimens. Some children report little limitation of activity or loss of friends, yet describe considerably less time spent with, and connection to, other children than their healthy peers.

Young children describe striving to maintain feelings of normality among their peers, and therefore avoid both spontaneous disclosure of their disease and response to their peers' direct questions about school absences. Children also report particular embarrassment about the need to administer eye drops in social settings; many describe skipping medications in such situations. Some children report that their parents help them problem-solve around this possibility, by discussing when and how they can administer eye drops discreetly at social events. The need for trips to the school nurse's office for eye drops or other medications may also be embarrassing for children who do not want to share details of their condition with peers. Some parents are not aware of their child's concerns, and others feel that their child should simply overcome them.

Although children with uveitis report that they generally do not talk with peers about their eye conditions (many not disclosing their conditions to anyone outside the family), some are aware of another child who also has uveitis, and feel that talking with someone who is dealing the same medical condition is helpful. With permission of all parties, we have

introduced selected patients to one another, and these patients have generally reported that their interactions are valuable. As children reach high school and early adulthood, they report beginning to disclose their disease to selected peers more openly, as they realize that peers also struggle with a variety of medical or other issues. Older children who have disclosed their condition to friends have generally found them to be supportive. This observation echoes a finding in the broader pediatric literature that perceived peer support predicts positive adjustment to chronic illness.¹⁶

Psychological Well-Being

Both patients and parents speak of difficult emotions that children experience because of uveitis and its treatment, including sadness, anxiety, and anger about attending multiple medical appointments, eye drops, and injections or infusions. Such distress can interfere with a child's willingness to participate in, and adhere to, medical treatments.

A common index of adjustment to chronic illness is the presence of psychological disorders. We have observed that some children experience marked distress while in the clinic and when discussing their experiences with uveitis, but it is unclear whether the observed distress is limited to disease-specific contexts, or whether it reflects more pervasive psychological problems. Furthermore, it is not clear which disease-related factors are most likely to influence children's psychological status. Interviewed parents mentioned that their children sometimes described distress related to disease experiences, but they did not observe emotional difficulties that permeated all areas of the children's lives.

The literature includes mixed findings about the vulnerability of children with chronic disease to psychological problems. Several studies of children with JIA suggest that their psychological functioning does not differ from that of healthy children.¹⁷⁻¹⁹ In contrast, other studies report that some with more severe JIA (and other rheumatic conditions) have a higher risk of psychological problems²⁰ and impaired quality of life.²¹ Studies of children with JIA have not isolated the contribution of associated uveitis, if any, to patients' psychological problems, but it is reasonable to assume that it would increase the risk of experiencing anxiety or depression. Parallels are found in other chronic diseases of childhood; for example, a higher risk of depression occurs among insulin-dependent diabetics with the most severe disease, based on poor glycemic control and more hospitalizations.²² Children with diabetes mellitus also show modest increases in depressive symptoms over the first 6 years after diagnosis, and girls evidence increases in anxiety as well.²² As with diabetes mellitus, the course of uveitis is experienced as unpredictable and difficult to control, and thus, children with more severe uveitis may be similarly at increased risk for elevated psychological distress and co-morbid psychological disorders.

Development of Autonomy

The literature on chronic illnesses of childhood frequently notes the difficulty of making the transition to independent adulthood. Approximately half of young adults with JIA do not successfully transfer their care from a pediatric rheumatologist to an adult rheumatologist.²³ Many children with chronic disease report especially close relationships with their parents and high levels of parental involvement.²⁴ Although emotional intimacy and parental

involvement can be positive forces, such influences can have unintended negative impact, if they impede a child's development of autonomy or intimate peer relationships.²⁵ In particular, children with chronic diseases are vulnerable to being unable to manage disease when parents are no longer present, particularly if they lack a history of communicating with physicians, managing medications, or recognizing changes in disease status.²⁴ On the other hand, patients may develop their own ideas about treatment as they get older and begin to receive information from sources outside the home, which could lead to conflict with parents who retain a major role in the health care of their adult offspring.

Children with chronic anterior uveitis may also transition to adulthood at a slower-than-average pace, with regard to medical management and independence from parents. Some adults in their 20s who have had chronic anterior uveitis since childhood continue to attend appointments accompanied by a parent. While young adults with low vision may need to rely on parents for transportation, even some with good vision do not pursue drivers' licenses or other independent forms of transportation, because they are accustomed to relying on others.

Young adults may continue living at home, if they rely on parents for management of medical appointments and treatment, for interactions with insurance companies, and for transportation. Some report embarrassment about their reliance on parents, particularly for transportation, while others appear content with the situation, neither seeing a necessity to become involved in their own medical care at this stage in their lives, nor predicting when they will assume that responsibility. We find that some families do not discuss this transition or prepare for it.

PARENTS' EXPERIENCE OF THEIR CHILDREN'S UVEITIS

Parents confront challenges in domains similar to those experienced by children as patients (Table 2).

Medical Challenges

Assuming responsibility for a child's medical care, including the need to deal with insurance companies, can dominate family life, especially at times when uveitis is not adequately controlled. As one mother stated, "Everything else in life takes a back seat". A father mentioned having "doctor's appointment fatigue." Decisions regarding the extent to which children will participate in their own care can also be stressful.

Vocational and Avocational Challenges

Because of the time needed to manage a child's disease, partial or complete withdrawal from career or other pursuits may be necessary for some parents. Especially in the case of reduced employment, financial consequences can loom large. Parents' involvement may extend to the child's academic domain. They may need to talk with school personnel to ensure adherence to treatment regimens, and some go themselves to the school to administer medications each day. Parents may also need to interact with school personnel to insure that their children receive accommodations to see the board in class or receive other forms of

help required by visual or physical limitations. Some report that requisite changes are obtained only after speaking repeatedly with teachers and administrators.

Interpersonal Relationships

Time devoted to managing medical issues can restrict a parent's energy for maintaining relationships with others. Couples may encounter interpersonal problems. They may perceive inequities in the responsibility for managing medical issues; they may have little time for focusing on their relationship as a couple; they may experience financial hardships; or they may disagree on what is best for their child. Nevertheless, some parents report that relationships with their partners are unaffected or actually strengthened through their shared responsibility for medical care. They emphasize the importance of engaging in a "team effort." In contrast, single parents can encounter heightened problems, unless they have strong social and material resources outside the home.

Parents may struggle to balance attention to the affected child and other offspring. Studies have shown a small, but significant, negative effect of chronic health conditions on psychological functioning of siblings.²⁶ Older siblings and siblings of children with more threatening and intrusive chronic health conditions were at highest risk.²⁶ We generally would not see such effects during patient care activities, and it was not a concern raised in our interviews, but it is a subject worthy of future consideration and study, specifically with regard to chronic anterior uveitis.

Relationships with friends can also be affected. Some parents report a fear of burdening those around them, if they talk about disease or request help, yet many express a desire to talk with other parents of children with uveitis. Some have done so and report psychological benefits, such as feeling less isolated, and pragmatic benefits, such as sharing tips about effective strategies for disease management.

Psychological Well-Being

Managing a child's disease can exact a considerable psychological toll on parents. For example, the need to palliate a child's nausea and vomiting the day after a methotrexate injection can be wrenching for parents, with substantial impact on a family's social life. Traveling long distances to attend frequent appointments for specialized care requires stamina. Even with close medical attention, chronic anterior uveitis can have an uneven course, and some parents speak of relentlessly "paddling upstream", as they live from one appointment to the next, hoping for good news about markers of inflammation or control of intraocular pressure. Many hope that the disease will simply remit, and only come to a difficult realization, months or years after the diagnosis, that the disease is indeed chronic.

Parents express a range of emotions about their children's conditions, including sadness and difficulty accepting the loss of a healthy childhood; anger at the injustice of a diagnosis; or a sense of isolation, if they know of no other family contending with uveitis. Uncertainty and fear about the future is common. At the same time, parents may speak of their admiration for an affected child, taking pride in how she or he copes with the disease. One parent described being "very proud and very scared at the same time." Commenting on the shifting nature of the disease and its nearly constant demands, a mother described "twice-a-month

meltdowns”, followed by support from a family member that allowed her to muster the energy to carry on.

Parents wish to encourage independence in their children, yet they maintain an acute awareness of the potentially grave consequences of unmanaged disease. One parent commented, “You do whatever you have to do to make sure your child gets care”, but as patients approach late adolescence, such intentions can evolve into an “internal struggle to let your child fly on her own.” Among mothers of children with JIA, a highly rated stressor is concern about being overly protective.²⁷ Research shows that parents’ psychological well-being influences children’s adjustment to chronic disease. Among children with rheumatic disease, Timko and associates found that maternal psychological characteristics, including a high sense of mastery over a child’s condition and low depression, are related to more positive adjustment of ill children.²⁰

DISCUSSION

Ophthalmologist readers of this article might wonder why many of the profound challenges described herein are not more obvious in their clinical interactions with patients and parents. Several factors may account for the relative invisibility of problems in routine encounters. Although the period immediately following diagnosis requires major adjustments, families often report that it is not until several years later that they feel the cumulative effect of various stressors, and come to understand that the burdens of managing uveitis can be unremitting. Given the relatively short time physicians spend with their patients, parents typically prioritize disease symptoms and treatment in their comments, as do physicians. Furthermore, parents vary in their perception of the appropriateness of raising issues about psychosocial adjustment with the ophthalmologist, or how welcome such comments might be, and they may be reticent to disclose difficulties in adhering to treatment recommendations.

Our impressions from working with children who have chronic anterior uveitis are reinforced by consistent themes across our interviews and by the work of others who have characterized the experiences of children with uveitis and their caregivers.¹¹ Our impressions include the following. Uveitis can be a greater focus of concern than concurrent systemic disease, and stressors extend beyond loss of vision or fear of blindness; in fact, many effects of uveitis are similar to those of other chronic diseases, and uveitis can impact quality of life even when there is no vision loss. Many stressors are related to the management of disease: the need for topical medications, which can be unpleasant; the injection or infusion of immunosuppressive medications, which can be painful and time-consuming; frequent trips to the ophthalmologist; and the experience of difficult and sometimes frightening examinations. Coping with these stressors varies markedly among different patients and families.

Health-related quality of life is impaired in multiple domains (physical, emotional, social, school) among children and adolescents with various chronic diseases, when compared to healthy controls.²¹ In many ways, the experiences of children with chronic anterior uveitis recall challenges encountered by children with insulin-dependent diabetes mellitus, in that

uveitis usually requires daily treatment and frequent medical appointments, and has the potential for serious long-term complications. There are sufficient parallels to suggest that children with uveitis face many of the same academic, interpersonal, and developmental challenges that have been well documented for other chronic disease populations.^{16,25} Children with both chronic anterior uveitis and JIA deal with the concurrent management of eye and joint disease, but it has been our impression that for many, the eye disease has a greater impact than joint disease on the patients' well-being, although research is needed to confirm that observation.

Parents can underestimate their child's quality of life.²⁸ Their perceptions may be influenced by their own quality of life,²⁹ as well as their anticipation of potential long-term consequences of the disease. Nevertheless, a capacity to maintain sound physical and psychosocial functioning, and even the ability to thrive, is evident in many families who experience chronic disease.³⁰ We have observed abundant examples of resilience in children and families, despite living with chronic anterior uveitis. Children who have never known life without uveitis often strive to make the best of their experiences.

We have observed that the ways parents cope with disease vary widely, as do their attempts to aid their children in adaptive coping. Our general impression is that parents most effectively help their child when they actively distinguish between the pliable and the intractable aspects of a disease, and focus coping efforts on altering controllable factors. While acknowledging some limitations imposed by the disease, parents can help children focus on what they, in fact, can do in life, rather than on their limitations. Research demonstrates that such coping responses are adaptive across many chronic illnesses of childhood, whereas coping attempts aimed at changing an immutable stressor, ruminating about the stressor, or avoiding the stressor do not bode well for adjustment.³¹ Furthermore, adopting a shared approach to coping can promote optimal functioning; for example, families of adolescent diabetics who view the disease as a shared responsibility between patient and parent experience better psychological health, self-care behaviors, and metabolic control over time.³²

Although informative, our findings have limitations. Impressions are based only on families with access to major medical centers and specialized care. An attempt was made to identify a pool of patients of various ages and experiences for interviews, but we cannot rule out selection bias. Families interviewed were those willing to speak with a psychologist, and may represent parents who are more aware of the health challenges they face than those in the general population with affected children.

Directions for Research

The themes mentioned above raise a number of possibilities for research on psychosocial factors related to uveitis. Exploratory interviews and clinical impressions are instructive, but are subject to bias and need to be validated further. This Perspective also describes several specific issues for which there is no information currently. Questions remain regarding the prevalence of uveitis-associated psychosocial difficulties and their determinants; the potential for ophthalmologists to promote adaptation; and how best to help families affected by uveitis. Research is needed on factors that affect families' disease management, and their

abilities to maintain accurate knowledge of frequent changes in drug regimens; a crucial need is research on approaches to bolster adherence. Neither information about adherence to ophthalmic treatments among adults nor information about general medical adherence among children with other diseases capture the nuanced combination of systemic and topical treatments required by those children with uveitis, yet an electronic literature search revealed no measures of adherence to treatment of eye-related diseases in children. In addition, the potential impact of uveitis during childhood on later quality of life in adulthood should be investigated.⁹

In response to the many calls for use of patient-reported outcomes in clinical research,³³ studies should examine the influence of uveitis and its treatment on children's quality of life using developmentally appropriate measures. Several child-specific measures of visual function are available, but they are not uveitis-specific.^{34–37} Measures to address the broader impact of uveitis need to be age-specific; deal with non-vision issues; and involve family members. The NEI-VFQ has been used widely to assess the impact of vision loss on quality of life in adults, but it includes questions that are not applicable to children and may not focus on concerns most relevant to them.³⁸ The EYE-Q (Effects of Youngsters' Eyesight on Quality of life) measures vision-related function and quality of life for children with uveitis who are 5–18 years of age.^{6,13,14} Worse EYE-Q scores are significantly related to lower visual acuity and bilateral ophthalmic disease.^{13,14} EYE-Q scores were significantly related to PedsQL scores, but the correlation was only moderate ($r=0.43$, $p<0.001$).¹⁴ The PedsQL does not show variation based on visual acuity, underlining the importance of a vision-specific quality of life indicator.¹⁴

Because stressors other than vision can have a major impact on a child's quality of life, it will be appropriate to include multiple measures in future research, with appropriate controls, which may help to identify changes in quality of life even among children with uveitis who retain excellent vision. In addition to the PedsQL, there are instruments that assess anxiety and depression in children and their medical transition readiness.^{39,40} Research is also needed on strategies for enhancing families' adjustment to uveitis. Facilitating contact with other patients and families dealing with uveitis, either in a professional setting or in a more informal context is thought to be beneficial in this respect, but it merits investigation.

Recommendations for Clinical Practice

Even without definitive research, ophthalmologists can be aware of patient reactions to chronic disease and its treatment, and to the impact of a child's disease on family functioning. Psychosocial impediments to optimal disease management provide a compelling reason for ophthalmologists to arm themselves with the requisite skills and referral sources to deal with these issues. Such skills may be as simple as taking an extra moment to express compassion or to normalize patients' and parents' experiences. They can offer patients the opportunity to discuss their frustrations, and they can address patients' specific concerns, helping them to develop active coping mechanisms, which may improve adherence to treatment regimens, resulting in better outcomes.

Ophthalmologists should begin to include young patients in discussions about their diseases. Age-appropriate explanations of disease mechanisms and some questions and instructions about disease management can be directed toward the child rather than solely toward the parents. Parents should be encouraged to involve their children incrementally in their own medical care, at levels appropriate for their ages and levels of maturity. From a practical standpoint, ophthalmologists can establish efficient procedures in their offices to document absences from school and to provide explanation of treatment regimens for school personnel. Physicians can also assist in the transition of older adolescents to independence in various ways: through referral to experienced colleagues, when patients relocate geographically for college; by timely transfer of medical records to new providers; and by encouraging adherence to treatment regimens and timely attention to signs or symptoms of disease exacerbation.

Many children and parents maneuver the complex problems associated with chronic anterior uveitis successfully, but for some families, the challenges are profound. Ophthalmologists are ideally positioned to raise awareness of such issues with families. Because of the need for frequent eye examinations, they often have more contact with a patient than her or his pediatric rheumatologist or even the primary care provider. Nevertheless, ophthalmologists may vary in their levels of comfort in addressing psychosocial issues directly with families. If such intervention is outside the scope of a physician's availabilities or expertise, referral to a clinical health psychologist, or to other professionals who are well-versed in the effects of chronic illness, is warranted.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

Funding: Supported by the Arthritis Foundation – Pacific Region and Mr. Al Pacino through gifts to the UCLA Stein Eye Institute Fund for Children with Uveitis (Dr. Holland); by the UCLA Stein Eye Institute Maggi Kelly Vision Fund (Dr. Holland); by the Skirball Foundation, New York, NY (Dr. Holland); by Award Number K23EY021760 from the National Eye Institute (Dr. Angeles-Han), the Rheumatology Research Foundation, Atlanta, Georgia (Dr. Angeles-Han), and the Cincinnati Children's Hospital Medical Center Research Innovation and Pilot Fund (Dr. Angeles-Han).

References

1. Holland GN, Stiehm ER. Special considerations in the evaluation and management of uveitis in children. *Am J Ophthalmol.* 2003; 135(6):867–878. [PubMed: 12788128]
2. Angeles-Han ST, Pelajo CF, Vogler LB, et al. Risk markers of juvenile idiopathic arthritis-associated uveitis in the Childhood Arthritis and Rheumatology Research Alliance (CARRA) Registry. *J Rheumatol.* 2013; 40(12):2088–2096. [PubMed: 24187099]
3. Angeles-Han ST, Yeh S, Vogler LB. Updates on the risk markers and outcomes of severe juvenile idiopathic arthritis-associated uveitis. *Int J Clin Rheumatol.* 2013; 8(1)doi: 10.2217/ijr.12.83
4. Holland GN, Denove CS, Yu F. Chronic anterior uveitis in children: clinical characteristics and complications. *Am J Ophthalmol.* 2009; 147(4):667–678. e665. [PubMed: 19195641]
5. Gregory AC 2nd, Kempen JH, Daniel E, et al. Risk factors for loss of visual acuity among patients with uveitis associated with juvenile idiopathic arthritis: the Systemic Immunosuppressive Therapy for Eye Diseases Study. *Ophthalmology.* 2013; 120(1):186–192. [PubMed: 23062650]

6. Angeles-Han ST, Griffin KW, Lehman TJ, et al. The importance of visual function in the quality of life of children with uveitis. *J AAPOS*. 2010; 14(2):163–168. [PubMed: 20236847]
7. Maca SM, Amirian A, Prause C, et al. Understanding the impact of uveitis on health-related quality of life in adolescents. *Acta Ophthalmol*. 2013; 91(3):e219–224. [PubMed: 23280117]
8. Angeles-Han ST. Quality-of-life metrics in pediatric uveitis. *Int Ophthalmol Clin*. 2015; 55(2):93–101. [PubMed: 25730622]
9. Haasnoot AJW, Sint Jago NFM, Tekstra J, de Boer JH. Impact of uveitis on quality of life in adult patients with juvenile idiopathic arthritis. *Arthritis Care Res (Hoboken)*. 2017; 69(12):1895–1902. [PubMed: 28217950]
10. Decarlo DK, McGwin G Jr, Bixler ML, Wallander J, Owsley C. Impact of pediatric vision impairment on daily life: results of focus groups. *Optom Vis Sci*. 2012; 89(9):1409–1416. [PubMed: 22863790]
11. Sen ES, Morgan MJ, MacLeod R, et al. Cross sectional, qualitative thematic analysis of patient perspectives of disease impact in juvenile idiopathic arthritis-associated uveitis. *Pediatr Rheumatol Online J*. 2017; 15(1):58. [PubMed: 28778202]
12. Miserocchi E, Modorati G, Mosconi P, Colucci A, Bandello F. Quality of life in patients with uveitis on chronic systemic immunosuppressive treatment. *Ocul Immunol Inflamm*. 2010; 18(4):297–304. [PubMed: 20482406]
13. Angeles-Han ST, Griffin KW, Harrison MJ, et al. Development of a vision-related quality of life instrument for children ages 8–18 years for use in juvenile idiopathic arthritis-associated uveitis. *Arthritis Care Res (Hoboken)*. 2011; 63(9):1254–1261. [PubMed: 21678564]
14. Angeles-Han ST, Yeh S, McCracken C, et al. Using the Effects of Youngsters' Eyesight on Quality of Life Questionnaire to measure visual outcomes in children with uveitis. *Arthritis Care Res (Hoboken)*. 2015; 67(11):1513–1520. [PubMed: 26037544]
15. Varni JW, Seid M, Kurtin PS. PedsQL 4.0: reliability and validity of the Pediatric Quality of Life Inventory version 4.0 generic core scales in healthy and patient populations. *Med Care*. 2001; 39(8):800–812. [PubMed: 11468499]
16. Suris JC, Michaud PA, Viner R. The adolescent with a chronic condition. Part I: developmental issues. *Arch Dis Child*. 2004; 89(10):938–942. [PubMed: 15383438]
17. Huygen AC, Kuis W, Sinnema G. Psychological, behavioural, and social adjustment in children and adolescents with juvenile chronic arthritis. *Ann Rheum Dis*. 2000; 59(4):276–282. [PubMed: 10733474]
18. Feldmann R, Weglage J, Roth J, Foell D, Frosch M. Systemic juvenile rheumatoid arthritis: cognitive function and social adjustment. *Ann Neurol*. 2005; 58(4):605–609. [PubMed: 16178013]
19. Ding T, Hall A, Jacobs K, David J. Psychological functioning of children and adolescents with juvenile idiopathic arthritis is related to physical disability but not to disease status. *Rheumatology (Oxford)*. 2008; 47(5):660–664. [PubMed: 18356173]
20. Timko C, Stovel KW, Moos RH, Miller JJ III. A longitudinal study of risk and resistance factors among children with juvenile rheumatic disease. *J Clin Child Psych*. 1992; 21(2):132–142.
21. Varni JW, Limbers CA, Burwinkle TM. Impaired health-related quality of life in children and adolescents with chronic conditions: a comparative analysis of 10 disease clusters and 33 disease categories/severities utilizing the PedsQL 4.0 Generic Core Scales. *Health Qual Life Outcomes*. 2007; 5:43. [PubMed: 17634123]
22. Kovacs M, Iyengar S, Goldston D, et al. Psychological functioning of children with insulin-dependent diabetes mellitus: a longitudinal study. *J Pediatr Psychol*. 1990; 15(5):619–632. [PubMed: 2283571]
23. Hazel E, Zhang X, Duffy CM, Campillo S. High rates of unsuccessful transfer to adult care among young adults with juvenile idiopathic arthritis. *Pediatr Rheumatol Online J*. 2010; 8:2. [PubMed: 20148143]
24. Holmbeck GN, Johnson SZ, Wills KE, et al. Observed and perceived parental overprotection in relation to psychosocial adjustment in preadolescents with a physical disability: the mediational role of behavioral autonomy. *J Consult Clin Psychol*. 2002; 70(1):96–110. [PubMed: 11860060]
25. Williams PG, Holmbeck GN, Greenley RN. Adolescent health psychology. *J Consult Clin Psychol*. 2002; 70(3):828–842. [PubMed: 12090386]

26. Vermaes IP, van Susante AM, van Bakel HJ. Psychological functioning of siblings in families of children with chronic health conditions: a meta-analysis. *J Pediatr Psychol.* 2012; 37(2):166–184. [PubMed: 21994420]
27. Barlow JH, Wright CC, Shaw KL, Luqmani R, Wyness IJ. Maternal stressors, maternal wellbeing and children's wellbeing in the context of juvenile idiopathic arthritis. *Early Child Dev Care.* 2002; 172(1):89–98.
28. Upton P, Lawford J, Eiser C. Parent-child agreement across child health-related quality of life instruments: a review of the literature. *Qual Life Res.* 2008; 17(6):895–913. [PubMed: 18521721]
29. Cremeens J, Eiser C, Blades M. Factors influencing agreement between child self-report and parent proxy-reports on the Pediatric Quality of Life Inventory 4.0 (PedsQL) generic core scales. *Health Qual Life Outcomes.* 2006; 4:58. [PubMed: 16942613]
30. Horton TV, Wallander JL. Hope and social support as resilience factors against psychological mothers who care for children with chronic physical conditions. *Rehabil Psychol.* 2001; 46(4): 382–399.
31. Compas BE, Jaser SS, Dunn MJ, Rodriguez EM. Coping with chronic illness in childhood and adolescence. *Annu Rev Clin Psychol.* 2012; 8:455–480. [PubMed: 22224836]
32. Helgeson VS, Reynolds KA, Siminerio L, Escobar O, Becker D. Parent and adolescent distribution of responsibility for diabetes self-care: links to health outcomes. *J Pediatr Psychol.* 2008; 33(5): 497–508. [PubMed: 17848390]
33. Deshpande PR, Rajan S, Sudeepthi BL, Abdul Nazir CP. Patient-reported outcomes: A new era in clinical research. *Perspect Clin Res.* 2011; 2(4):137–144. [PubMed: 22145124]
34. Gothwal VK, Lovie-Kitchin JE, Nutheti R. The development of the LV Prasad-Functional Vision Questionnaire: a measure of functional vision performance of visually impaired children. *Invest Ophthalmol Vis Sci.* 2003; 44(9):4131–4139. [PubMed: 12939337]
35. Birch EE, Cheng CS, Felius J. Validity and reliability of the Children's Visual Function Questionnaire (CVFQ). *J AAPOS.* 2007; 11(5):473–479. [PubMed: 17512228]
36. Khadka J, Ryan B, Margrain TH, Court H, Woodhouse JM. Development of the 25-item Cardiff Visual Ability Questionnaire for Children (CVAQC). *Br J Ophthalmol.* 2010; 94(6):730–735. [PubMed: 20508047]
37. Cochrane GM, Marella M, Keeffe JE, Lamoureux EL. The Impact of Vision Impairment for Children (IVI_C): validation of a vision-specific pediatric quality-of-life questionnaire using Rasch analysis. *Invest Ophthalmol Vis Sci.* 2011; 52(3):1632–1640. [PubMed: 21178144]
38. Mangione CM, Lee PP, Pitts J, et al. Psychometric properties of the National Eye Institute Visual Function Questionnaire (NEI-VFQ). NEI-VFQ Field Test Investigators. *Arch Ophthalmol.* 1998; 116(11):1496–1504. [PubMed: 9823352]
39. Chorpita BF, Yim L, Moffitt C, Umemoto LA, Francis SE. Assessment of symptoms of DSM-IV anxiety and depression in children: a revised child anxiety and depression scale. *Behav Res Ther.* 2000; 38(8):835–855. [PubMed: 10937431]
40. Sawicki GS, Lukens-Bull K, Yin X, et al. Measuring the transition readiness of youth with special healthcare needs: validation of the TRAQ--Transition Readiness Assessment Questionnaire. *J Pediatr Psychol.* 2011; 36(2):160–171. [PubMed: 20040605]

TABLE 1

Summary of Potential Psychosocial Challenges Experienced by Children with Chronic Anterior Uveitis.

| |
|--|
| A. Medical Challenges. |
| 1. Maintenance of visual acuity and compensating for impaired vision. |
| 2. Management of complex and changing treatment regimens. |
| 3. Adherence to medical regimens. |
| 4. Negotiation of relative responsibility with parent for medical management. |
| B. Academic Competence. |
| 1. Interference of visual changes with ability to complete schoolwork. |
| 2. Absences due to treatment and regular ophthalmology appointments. |
| 3. Disruption of class time to visit nurse's office for eye drop administration. |
| C. Interpersonal Relationships and Recreational Activities. |
| 1. Limited participation in some activities because of impaired vision, treatment, and side effects of treatment. |
| 2. Uncertainty regarding disclosure to peers, and regarding peer reactions to disease status, if disclosed. |
| 3. Absence of peers with similar experiences. |
| 4. Embarrassment regarding appearance (e.g., patches, redness, strabismus) and reaction to taunting about appearance or disease by peers. |
| D. Psychological Well-being. |
| 1. Psychological distress related to uncertain course of disease or treatment. |
| 2. Negative emotions about unpleasant treatment side effects. |
| E. Development of Autonomy. |
| 1. Importance of disease control may lead to high levels of parental monitoring and involvement, even into adulthood. |
| 2. Complex, frequently changing regimens of corticosteroids and immunosuppressive medications may be difficult for children to understand and to remember. |
| 3. Interference in meeting developmental milestones due to disease complications or demands of treatment (e.g., driving, financial independence). |

Table 2

Summary of Potential Psychosocial Challenges Experienced by Parents of Children with Chronic Anterior Uveitis.

| |
|--|
| A. Medical Challenges. |
| 1. Increased time demands to attend medical appointments and provide care for children during school absences. |
| 2. Increased financial burden, to cover medical co-payments and treatments not covered by insurance. |
| 3. Negotiation of relative responsibility with child for medical management. |
| B. Vocational and Avocational Challenges. |
| 1. Reduction of or withdrawal from career or other pursuits. |
| 2. Financial consequences of reduction in employment. |
| C. Interpersonal Relationships. |
| 1. Increased attention to child may detract time and energy from other relationships. |
| 2. Negotiation of shared management of child's treatment with partner. |
| 3. Dealing with a child's concern about becoming a burden to family and friends. |
| 4. Potential for strengthened relationships through shared adversity. |
| 5. Prolonged parenting commitments due to a child's delayed achievement of autonomy. |
| D. Psychological Well-being. |
| 1. Uncertainty about offspring's current disease status, success of medical regimens, trajectory of disease. |
| 2. The need to help children cope with negative emotions related to disease-associated challenges can be emotionally taxing. |
| 3. Treatment demands may overwhelm parental psychological resources and deplete time for self-care. |
| 4. Potential for strengthening psychological resources through adversity. |