

## Patient-derived questionnaire items for patient-reported outcome measures in adults with strabismus

Sarah R. Hatt, DBO, David A. Leske, MS, and Jonathan M. Holmes, BM, BCh

**In a previous study we performed individual interviews of adults with strabismus and identified specific health-related quality of life concerns. In this present report we provide the entire list of specific questionnaire items that were created, one for each concern articulated in the interviews. Our aim in reporting this full list of 181 individual, patient-derived questionnaire items is to provide a resource to clinicians and researchers interested in developing patient-reported outcome measures for adults with strabismus.**

Patient-reported outcome measures are important for the assessment of health, well-being, and treatment effectiveness. We have previously reported the development of the Adult Strabismus (AS)-20 questionnaire, a patient-reported outcome measure evaluating health-related quality of life (HRQOL) in adults with strabismus.<sup>1,2</sup> In developing the AS-20 questionnaire, we created an initial list of all potential questionnaire items based on patient-derived concerns identified from individual patient interviews.<sup>1,3</sup> We have not previously reported this entire list of potential questionnaire items, and we now see the value of making these items available as a resource to clinicians and researchers. In the present report we provide the full list of candidate questionnaire items originally identified from individual interviews of adults with strabismus.

### Subjects and Methods

Approval for this study was obtained from the Mayo Clinic Institutional Review Board. All procedures and data collection were conducted in a manner compliant with the US Health Insurance Portability and Accountability Act of 1996, and all research procedures adhered to the tenets of the Declaration of Helsinki. Informed consent was obtained.

*Author affiliations: Department of Ophthalmology, Mayo Clinic, Rochester, Minnesota*  
*Study conducted at: Department of Ophthalmology, Mayo Clinic, Rochester, Minnesota.*  
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*Correspondence: Dr. Jonathan M. Holmes, BM, BCh, Ophthalmology E4, Mayo Clinic, Rochester, MN 55905 (email: holmes.jonathan@mayo.edu).*  
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As described previously,<sup>1,3</sup> 30 adults with strabismus (17 [57%] with diplopia and 13 [43%] without diplopia), across a range of diagnoses, were enrolled from a single strabismus practice (JMH). Each patient was interviewed using 11 open-ended questions in a semistructured format.<sup>3</sup> Each interview was audiorecorded and transcribed, and transcripts were reviewed independently by three investigators.<sup>3</sup>

Each of the three investigators independently identified specific concerns in each interview. For each patient, the three lists of concerns (one from each investigator) were combined, and duplicates were removed.<sup>3</sup> Concerns were then grouped by theme across all patients.

Two investigators reviewed the entire list of concerns, derived from all patient interviews and formulated a specific question for each specific concern. Questionnaire items were formulated as statements.

### Results

Across all individual patient interviews a total of 1,301 specific concerns were extracted that pertained to HRQOL.<sup>3</sup> From these concerns, after duplicates were removed, a total of 181 individual questionnaire items addressing specific HRQOL concerns were formulated.<sup>1</sup> This entire list of derived questionnaire items is provided in the eTable 1, grouped by theme.

### Discussion

We have made available the entire list of questionnaire items derived from individual interviews of adults with strabismus. Making this complete list of items available provides a resource to other care providers and researchers interested in developing patient-reported outcome measures.

In the development of patient-reported outcome measures it is important to utilize the patient's perspective when establishing the content of items.<sup>4,5</sup> This may be achieved using individual patient interviews or focus groups, and in the present study we elected to use individual patient interviews. By interviewing patients with a range of strabismus types and severities, and recording each specific concern we aimed to capture the full range of HRQOL issues affecting adults with strabismus.

The main purpose of making the complete list of potential questionnaire items publicly available is to provide other care providers and researchers with a resource to use in the development of other patient-reported outcome measures. One approach would be to use the statements as questions in an item bank,<sup>6,7</sup> initially administering all the items to a large group of patients, performing factor analysis or principal component analysis to establish unidimensional domains, and performing Rasch analysis to Rasch-calibrate each item. This process could then lead to computer-adaptive testing,<sup>6</sup> which would allow rapid administration of a few questions to provide a Rasch-calibrated score for each domain. Alternatively, short forms could be developed such as the AS-20 questionnaire.<sup>1</sup> The AS-20 has undergone Rasch analysis<sup>2</sup> and is now freely available online, with Rasch look-up scoring tables ([www.pedig.org](http://www.pedig.org)).

There are some limitations to our study. Although we interviewed a heterogeneous group of strabismus patients, it is possible we missed some concerns that may be specific to a particular subpopulation of adults with strabismus, or may be present in other cultures. Nevertheless, we included each concern even if it was only mentioned once.

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## Accommodative esotropia and Brown syndrome in a girl with recessive geleophysic dysplasia

Arif O. Khan, MD,<sup>a,b</sup> and Patrik Schatz, MD, PhD<sup>c,d</sup>

**Geleophysic dysplasia and Weill-Marchesani syndrome are acromelic dysplasias characterized by short stature, brachydactyly, and joint contractures. Recessive Weill-Marchesani syndrome typically includes spherophakia, but the ocular phenotype of recessive geleo-**

*Author affiliations:* <sup>a</sup>Eye Institute, Cleveland Clinic Abu Dhabi, Abu Dhabi, United Arab Emirates; <sup>b</sup>Department of Ophthalmology, Cleveland Clinic Lerner College of Medicine of Case Western University, Cleveland, Ohio; <sup>c</sup>Vitreo-retinal Division, King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia; <sup>d</sup>Department of Ophthalmology, Clinical Sciences, Skane County University Hospital, University of Lund, Lund, Sweden  
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Correspondence: Arif O. Khan, MD, Eye Institute, Cleveland Clinic Abu Dhabi, PO Box 112412, Abu Dhabi, United Arab Emirates (email: arif.khan@mssm.edu).  
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**physic dysplasia is not well defined. We describe the ocular phenotype of a girl with genetically confirmed recessive geleophysic dysplasia (biallelic *ADAMTSL2* mutations). Features included high corneal astigmatism, accommodative esotropia, unilateral Brown syndrome, and no evidence for zonular disease at 12 years of age.**

## Case Report

A 10-year-old girl previously diagnosed with familial geleophysic dysplasia and confirmed to harbor compound heterozygous mutations in the gene *ADAMTSL2* (c.338G>T/IVS5-89G>A) that segregated with the phenotype was referred for ophthalmic examination because of eye misalignment. The girl, born to first cousins, and her 2 affected sisters (not available for examination) were the subject of a prior report highlighting cardiac involvement in their disease.<sup>1</sup> Consistent with her diagnosis of geleophysic dysplasia, she had short stature, multiple infantile respiratory infections, joint contractures, and static cardiac valvular disease.<sup>1</sup> On examination, her affect and social interaction seemed appropriate for age. She was small (height, 115.4 cm [3rd percentile, 122 cm], weight 20.5 kg [3rd percentile, 19 kg]), with acromelia. Her facies were notable for thin upper lip, smooth and wide philtrum, anteverted nares, hypertelorism, and broad nasal bridge. She did not exhibit an abnormal head position. Visual acuity (without correction) was 20/60 in each eye. At near there was an esotropia of approximately 35<sup>Δ</sup> and hypotropia of 10<sup>Δ</sup>. Versions were significant for -2 elevation in adduction in the left eye that did not improve with ductions, consistent with Brown syndrome (Figure 1). Slit-lamp examination was normal, including no evidence for spherophakia or ectopia lentis before and after pharmacological pupillary dilation. Cycloplegic refraction (cyclopentolate 1%) was +5.50 -3.50 × 180 in the right eye and +6.00 -4.00 × 180 in the left eye. Fundus examination was significant for small crowded optic nerve heads in both eyes. She was prescribed her full hyperopic correction.

On follow-up examination 3 months later, visual acuity with correction was 20/20 in each eye. With correction, at near was an esotropia of 14<sup>Δ</sup> and right hypotropia of 10<sup>Δ</sup> in primary position. At 12 years of age her refraction and ocular motility were unchanged. Ocular biometry (Zeiss IOLmaster) was as follows: keratometry 42.99 D/48.08 D @072 in the right eye and 43.55 D/48.28 D @107 in the left eye; anterior chamber depth was 3.01 mm in the right eye and 3.06 in the left eye; and axial length was 20.92 mm in the right eye and 20.45 mm in the left eye.

## Discussion

The ocular features of this child with recessive geleophysic dysplasia secondary to biallelic *ADAMTSL2* mutations were short eyes, high corneal astigmatism, and left Brown syndrome. There was no evidence for zonular disease at 12 years of age.

eTable 1. Full list of 181 questionnaire items derived from individual semistructured interviews of 30 adults with strabismus

No.	Topic	Question
1	Visual function- general	I have problems looking one direction or another
2		I see things in different places than they actually are
3		I have a hard time seeing what's in front of me
4		I have problems seeing fine details
5	General disability	I feel handicapped because of my eyes
6		I am unable to function normally because of my eyes
7		I feel that my eyes affect my quality of life
8		My eyes affect my independence
9	Adaptation	I avoid things that give me problems
10		My eyes limit what I can do
11		I try to hide my eyes from other people
12		I have to use extra caution when doing things
13	Compensatory body posture	I joke about my eyes to feel better around others
14		My eye condition makes me do things differently
15		I turn my head a certain way to see better
16		I have problems looking at things with my head straight
17	Monocular eye closure	I cover or close one eye to see things better
18	Self-help	I try to pull my eyes together
19		I try to correct my eyes
20		I have to blink my eyes to see things
21		I am unable to control my eyes
22	Concentration	I have a hard time concentrating
23		I stop doing things because my eyes make it difficult to concentrate
24		I do things slower than other people
25		I need to concentrate more than others
26	Bump into things	I run into walls
27		I bump into people
28	Busy/strange environments	I have problems finding new places
29		I have problems walking in crowds
30		I have problems in busy environments
31		My eyes are worse in busy environments
32	Depth perception	I have problems seeing things that move fast
33		I get distracted by things that move
34		I have problems with depth perception
35		I have problems setting things on the table or counter
36	Specific disability	I misjudge distances
37		I have a hard time determining if something is coming towards me
38		I have problems doing everyday tasks because of my eyes
39		Shopping is hard for me because of my eyes
40	Steps	I find it hard to do housework because of my eyes
41		I have problems using a mirror
42		It is hard to put a key in a lock
43		I have problems using a screwdriver and a screw
44	Walking	I avoid climbing ladders
45		I find it difficult to go up and down steps
46		I make extra steps when going up or down stairs
47		I step too high or low when going up stairs
48	Lighting	I avoid stairs
49		I fall off curbs
50		I have to take extra care when walking
51		I stumble when walking
52	Physical discomfort	I have to watch my steps
53		I have problems walking on uneven surfaces
54		My eyes are worse in bright sunlight
55		Changing lighting makes my eyes worse
56	Headaches	Fluorescent light bothers me
57		I am bothered by bright lights at night
58		My eyes hurt
59		I can feel my eye wandering
60	Eye fatigue	My eyes feel strained
61		My eyes give me headaches
62		My eyes get tired
63		I have to rest my eyes
64	Effect of fatigue	My eyes are worse when I'm tired

(Continued)

eTable 1. Full list of 181 questionnaire items derived from individual semistructured interviews of 30 adults with strabismus (*Continued*)

No.	Topic	Question
65	Back/neck pain	My neck hurts because of my eyes
66		My back hurts because of my eyes
67	Prisms	I am dependent on my glasses
68		I dislike having to wear prism glasses
69	Non-specific negative emotions	Living with my eye condition is very hard
70		I feel sad because of my eyes
71		I feel like crying because of my eyes
72		I feel emotionally drained because of my eyes
73		I feel stressed because of my eyes
74		I feel angry about my eyes
75	Annoying	I get annoyed because of my eyes
76		My eyes bother me
77	Anxiety	I worry about my eyes
78		I worry that my eyes will get worse
79		I am conscious of my eyes
80		I worry that I will mess up because of my eyes
81		I worry about what people will think about my eyes
82		I worry that surgery won't fix my eyes
83		I worry about going blind
84	Frustrating	People's comments about my eyes frustrate me
85		My eyes frustrate me
86	Jealous	I am jealous of people with straight eyes
87	Resignation	I have problems accepting my eye condition
88		I expect my eyes to get better <sup>a</sup>
89	Lack of support	People don't understand my eye condition
90		People think I make things up about my eye condition
91	Teasing	People insult me because of my eyes
92		People call me names because of my eyes
93		People tease me because of my eyes
94	Self-confidence	I don't feel confident because of my eyes
95		I feel normal <sup>a</sup>
96		Having normal eyes would make a big difference to my self confidence
97		I am more shy because of my eyes
98	Self-conscious	I am embarrassed by my eyes
99		I feel uncomfortable when people are looking at me
100		I wonder what people are thinking when they are looking at me
101		I am self-conscious about my eyes
102		I feel different from other people
103		I wonder if people notice my eyes
104	Self-esteem	I feel useless because of my eyes
105		I have low self-esteem because of my eyes
106		I feel inferior to others because of my eyes
107	Appearance to others	Other people stare at my eyes
108		People judge me because of my eyes
109		People don't think I'm normal because of my eyes
110		People comment about my eyes
111		People think I'm weird because of my eyes
112		I feel that people are thinking about my eyes even when they don't say anything
113	Appearance to self	I don't like the way my eyes look
114		I wear sunglasses to cover my eyes
115		I am conscious that my eyes are going in different directions
116		I dislike the way I look in pictures
117		I dislike the way my glasses look
118	Confidence of others	People treat me differently because of my eyes
119		People are uncomfortable around me because of my eyes
120		People don't have confidence in my abilities because of my eyes
121		People don't respect me because of my eyes
122		People don't give me opportunities because of my eyes
123	Eye contact	I find it hard to look people in the eye
124		I avoid eye contact
125		I look away when talking to people
126	Eye contact from others	People avoid looking at me
127		People ask who I am looking at
128		People don't know where I'm looking

*(Continued)*

eTable 1. Full list of 181 questionnaire items derived from individual semistructured interviews of 30 adults with strabismus (*Continued*)

No.	Topic	Question
129		People don't know which eye to look at
130		My eyes make other people feel uncomfortable
131	Social	My eyes affect my social life
132		I don't go out much because of my eyes
133		I avoid people because of my eyes
134		I have a hard time recognizing faces
135	Interpersonal relationships	People talk to me like I'm stupid
136		My eyes affect the way I talk to people
137		My eyes make it harder for me to make friends
138		People react differently to me because of my eyes
139	Meeting people	I avoid people of the opposite sex because of my eyes
140		I find it hard to initiate contact with people I don't know
141		It is hard for me to meet people
142	Hobbies	I can't enjoy my hobbies
143		I have difficulties watching TV
144		Watching a movie in the theater is hard for me
145		I can't do the things I like to do
146	Reading	I avoid reading because of my eyes
147		I have problems reading
148		I read slower than other people
149		My eyes hurt when I read
150		I need to take frequent breaks when reading
151	Sewing	I have a hard time sewing
152		I have a hard time sewing a button on a shirt
153		It is hard for me to thread a needle
154	Driving	I have problems with driving because of my eyes
155		I have to take extra care when driving because of my eyes
156		I avoid driving because of my eyes
157		I have a hard time parking the car because of my eyes
158		I worry about driving into things because of my eyes
159		I don't feel safe while driving because of my eyes
160		I can't tell how fast cars are moving
161		I have problems driving at night
162	Cooking	I have a hard time cooking
163		I have a hard time pouring liquids into a glass
164	Sports	It is difficult for me to play sports because of my eyes
165		I have problems catching a ball
166		I have problems hitting a ball with a bat
167		I have problems hitting a tennis ball
168		I have problems seeing things that move fast
169	Work	My eyes affect my ability to work
170		My eyes have prevented me going further in my job
171		I find it hard to get a good job because of my eyes
172		It takes me longer to do things than other people
173		My eyes have limited the type of job I can do
174		I need to take frequent breaks when working
175	Financial	My eye condition has affected me financially
176		My eyes affect my ability to earn money
177		I miss work because of my eyes
178		Paying for glasses has been a financial burden for me
179	Computer	I have problems looking at a computer screen
180		I have problems viewing a computer screen
181		I have to sit far away from a computer screen

<sup>a</sup>Items that would require reverse scoring.