Plate 1

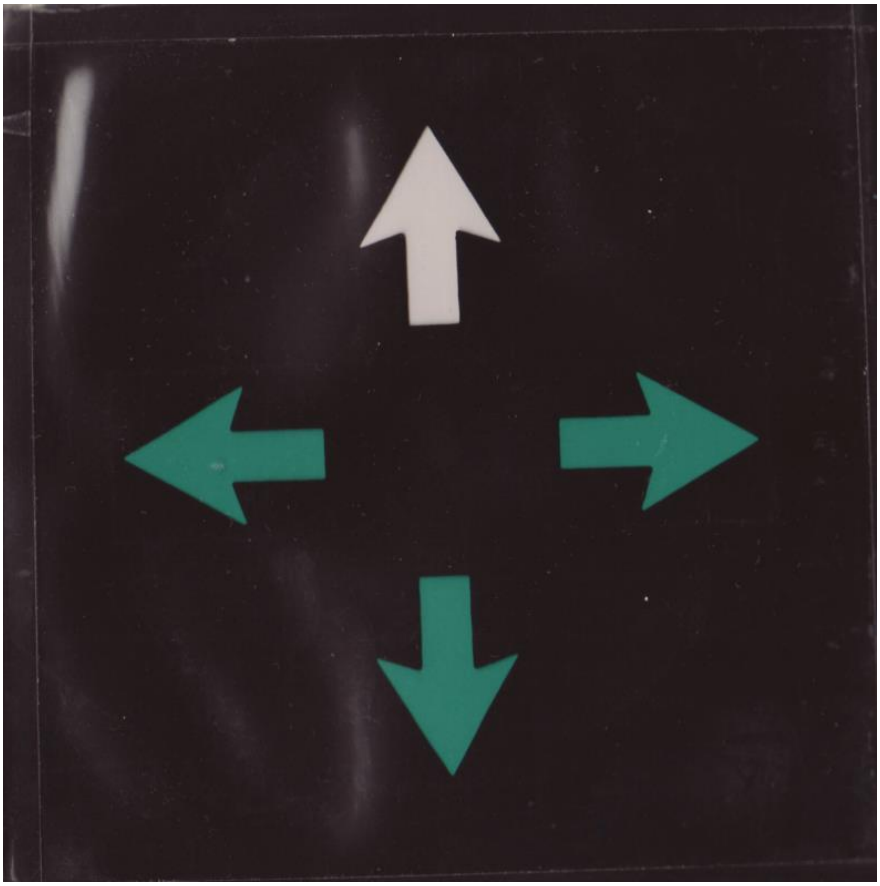


Plate 2

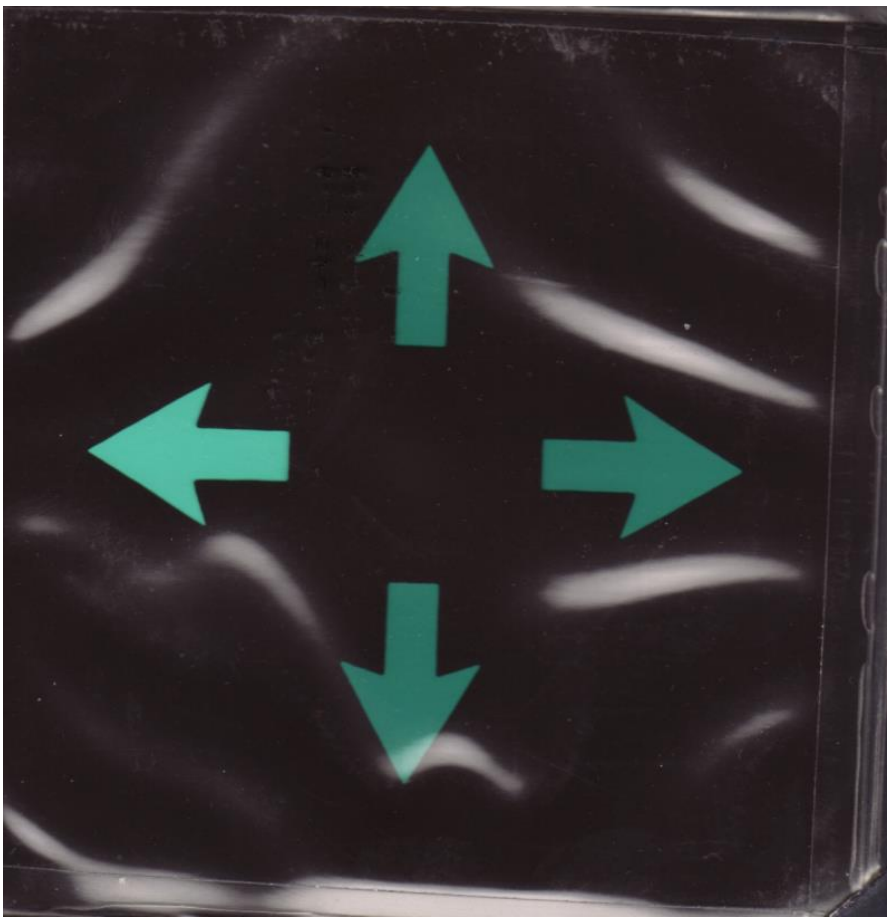


Plate 3

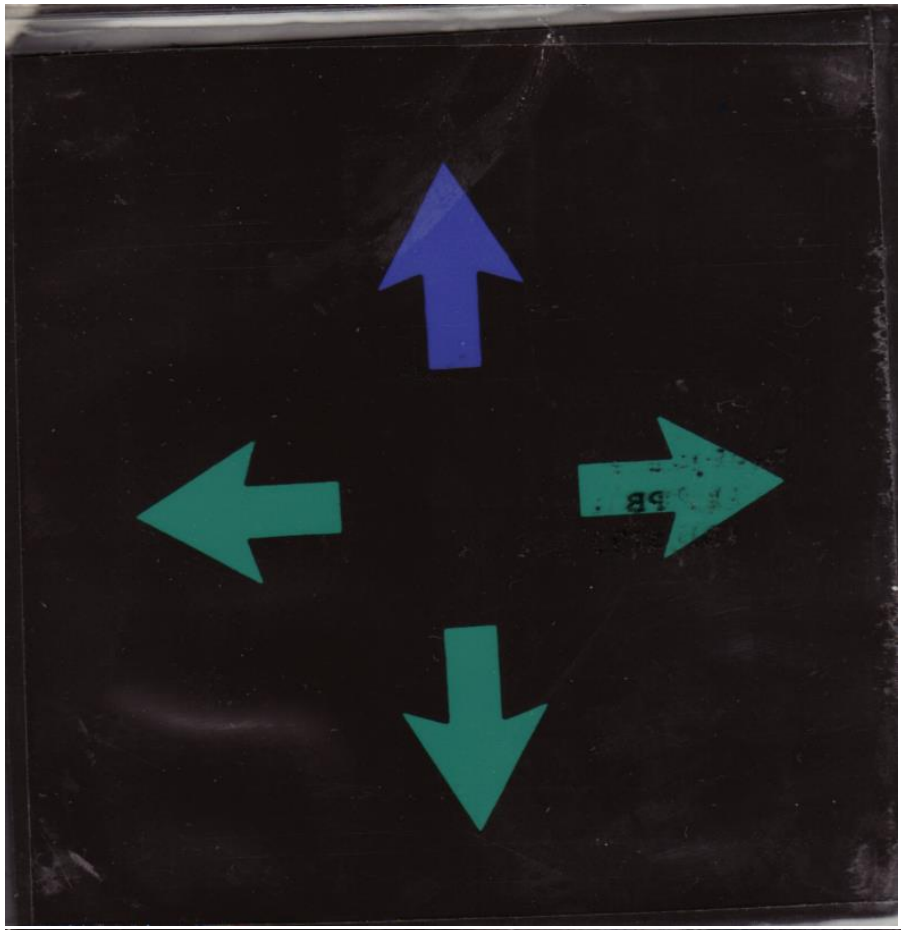


Plate 4

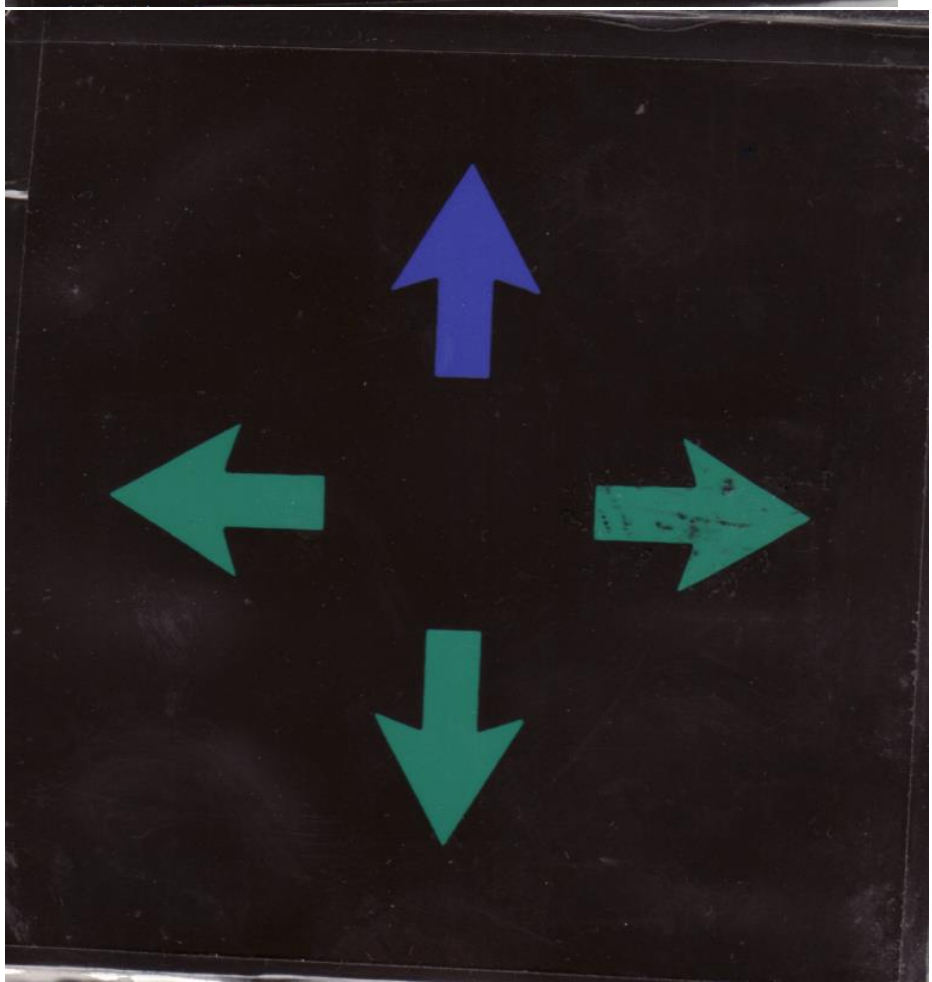


Plate 5

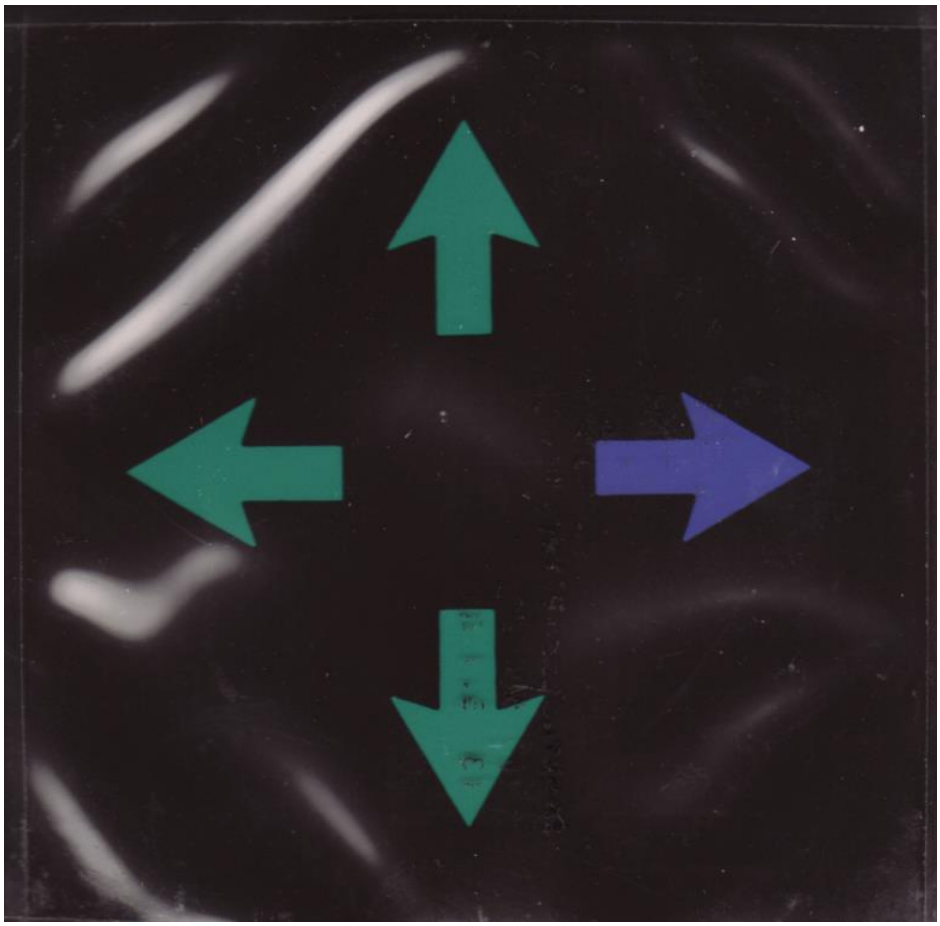
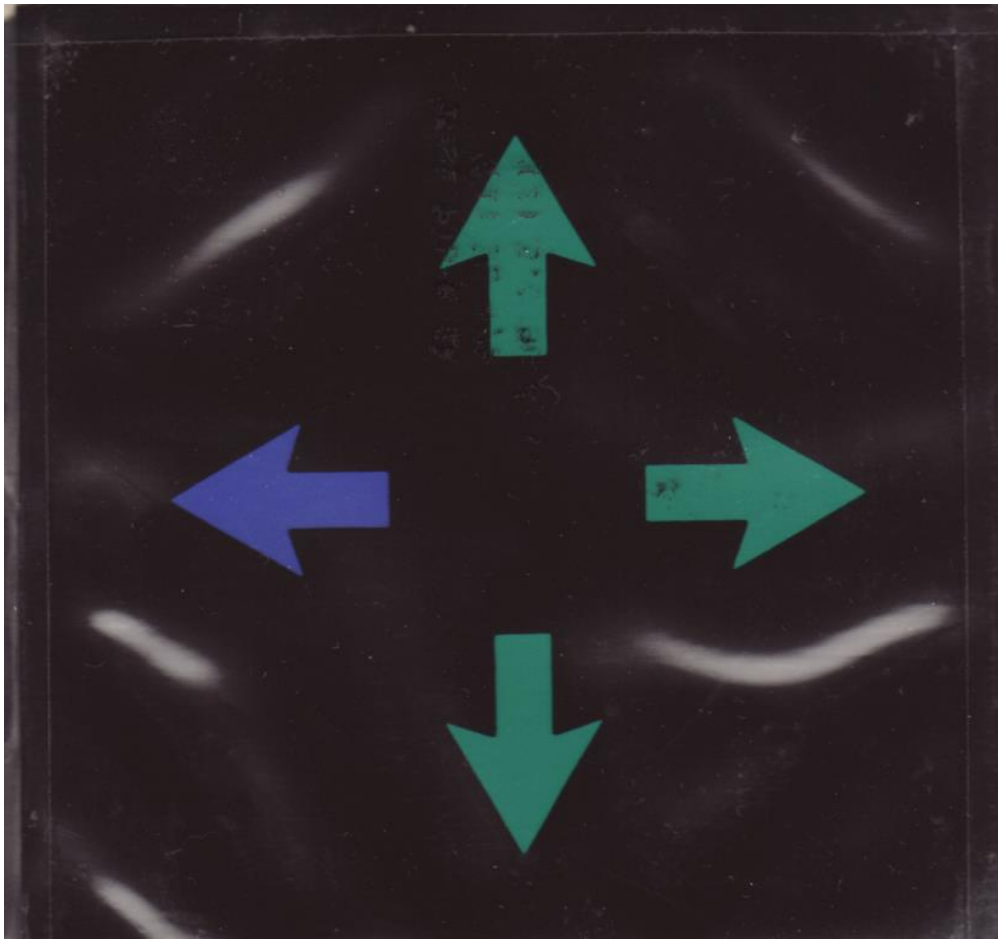


Plate 6



BLUE CONE MONOCHROMATISM PLATES

1. Description and clinical application.

This color vision test consists of six plates, including two instructional and four test plates. Patients must be able to identify the odd gray and odd light-green arrow on instructional plates 1 and 2 respectively before they are evaluated with the four test plates. Each test plate has three identical blue-green arrows and one purple-blue arrow; test plates differ from one another only with respect to the chroma of the purple-blue arrow. Young patients with blue cone monochromatism can easily distinguish the purple-blue arrow on all four test plates, whereas patients with rod monochromatism cannot distinguish the purple-blue arrow on all four plates. If a male has reduced visual acuity, normal rod electroretinograms, and 30 Hz cone electroretinograms reduced more than 97% below normal (i.e., less than 1.5 μV), this test can be used to determine whether his condition is an X-chromosome-linked one or an autosomal recessive one.

2. Patients eligible for this test.

Candidates for this test are males with photophobia and color-blindness, corrected visual acuity of about 20/200 in both eyes, nystagmus, clear ocular media, and a normal or nearly normal fundus appearance on ophthalmoscopic examination. Rod full-field electroretinograms must be normal, and cone full-field electroretinograms to 30 Hz white flicker must be less than 1.5 μV in amplitude.

3. Instructions for administering the test.

We recommend the following procedure for administering this test. The patient is seated and views the plates on a desk at a distance of about 40 cm. The patient wears any prescription lens (untinted) that is normally worn; one eye is patched. The room is darkened and the plates are then illuminated with a Macbeth lamp; the lamp provides a spectrum equivalent to standard illuminant C. The examiner says, "I am going to show you a series of pictures with arrows pointing in different directions. I will ask you to point to the arrow that looks different. Here is an example." The examiner then shows instructional plate 1 and points to the odd gray arrow. The examiner then removes this plate from the patient's view, randomly rotates it, and again places it in front of the patient, stating, "Let's see if you can pick out the arrow that is different." The patient can also be asked to point in the same direction as that of the odd arrow to confirm that he can see the arrows. This procedure is repeated several times until the examiner is certain that the patient can identify the odd arrow. The examiner then repeats this procedure with instructional plate 2, making six consecutive presentations of this plate, each preceded by random rotation of the plate out of the patient's view. If the patient can correctly identify the odd arrow on all six presentations, then it is clear ($P < .001$) that the patient is not selecting the odd arrow at random and therefore understands the test. If the patient cannot identify the arrow on plate 2, then he is re-instructed on plate 1 and then shown plate 2 again. Only if the patient can perform plate 2 can the subsequent testing be considered valid.

(continued)

BLUE CONE MONOCHROMATISM PLATES

Once the patient passes plate 2, the examiner states, "Now you know how to do the test. On the following plates, point to the arrow that is different. If you cannot see a difference, you can guess." The examiner then presents test plates 1 to 4, respectively, six consecutive times; before each presentation the examiner randomly rotates the plate out of the patient's view. The patient is given about 30 seconds to make each choice. After testing of the first eye is complete, the same sequence (including presentation of the instructional plates) is repeated on the second eye with the first eye patched.

To pass the test with each eye, the patient must correctly identify the odd (purple-blue) arrow on at least three of six presentations on each of the four test plates to ensure to the $P \leq .001$ level of confidence that he is not selecting the odd arrow by chance. We adopted this criterion for passing this test because if one assumes that the patient with rod monochromatism is selecting arrows at random, the probability (from the binomial distribution) that this individual will identify the correct arrow by chance on three out of six presentations of a given plate is 0.169. The probability of this occurring by chance on each of four plates is 0.0008 (that is, 0.169^4). Therefore, we chose the criterion of at least three correct selections out of six presentations on each of four test plates because this was the least stringent criterion with a probability of less than .001. Blue cone monochromats easily see all four test plates, whereas rod monochromats have great difficulty, even with the occasional test plate they can perform.