Congenital tritanopia without neuroretinal disease

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Recently the view that congenital tritanopia does not exist and that tritanopia is always secondary to ocular disease has been revived. It has been suggested that previous accounts of congenital tritanopia have not adequately excluded ocular disease and specifically that tritanopia thought to be congenital may in fact be secondary to dominantly inherited juvenile optic atrophy, a disease which often presents with signs sufficiently subtle as to go unnoticed by the patient and the examiner. This paper reports the investigation of six tritans from two families already documented in the literature as exhibiting congenital tritanopia. The investigation sought evidence of ocular disease and in particular evidence of dominantly inherited juvenile optic atrophy. No such evidence was found and it is concluded, that their tritanopia is not acquired. It seems that congenital tritanopia does exist and moreover shows distinct differences from tritanopia acquired secondary to dominantly inherited juvenile optic atrophy.

Key words: Tritanopia, tritanomaly, color vision, acquired color vision, color vision tests, human genetics, familial optic atrophy.

Inherited congenital tritanopia was first reported in the last quarter of the nineteenth century, but of the early works only that of Konig in 1897 received wide and lasting recognition. Konig examined nine cases of tritanopia secondary to ocular disease and drew up a relatively sophisticated model of the color vision defect. It is likely that the emphasis Konig’s work placed on the possible acquired nature of tritanopia led scientific opinion to doubt the existence of any other form. Certainly, the belief that congenital tritanopia was rare or even that it did not exist was common among and quite acceptable to many eminent visual scientists.

Parsons wrote that “cases of tritanopia are rare, and mostly due to disease.” In 1944, Pitt claimed “tritanopia is almost invariably due to disease” and he went on to suggest that even cases of tritanopia where disease could not be found were merely simulations of tritanopia caused by sclerosis of the crystalline lens or extremely dense macular pigmentation. This contemporary obsession with the effects of macular pigmentation led Walls and Mathews to state...
in its most extreme form the disease of "let's blame it on the macular pigment" attacks tritanopia. Although they decried the use of macular pigmentation as the whipping-boy for congenital tritanopia, Walls and Mathews in that same monograph stated "there is probably no such thing as congenital hereditary tritanopia" and "congenital tritanopia is, however, so extremely rare that it seems to be tacitly agreed that there must be no gene for it, so that the occasional case must have a nonhereditary basis."

It has been generally assumed that any doubt that inherited congenital tritanopia might exist was dispelled by the classic study of Wright in 1952; he confirmed 17 cases of tritanopia and carried out extensive laboratory measurements on seven of these. Subsequent studies by Kalmus, Sperling, Walls, Henry, Cole, and Nathan, Cole, Henry, and Nathan, and Schmidt reinforced and augmented Wright's findings.

Recently, however, the view that all cases of tritanopia are secondary to ocular disease has been revived by Krill, Smith, and Pokorny who have hypothesized that congenital tritanopia does not exist, and cases mistakenly reported as such in the past have actually been undiagnosed cases of dominantly inherited juvenile optic atrophy (DIJOA). They were led to this conclusion by their failure and that of some others to find a single case of congenital tritanopia, and because in their opinion, the presence of a causative ocular disease had not been adequately excluded in previous reports of that condition. They selected DIJOA as the likely disease with which congenital tritanopia has been confused because in addition to a tritan dyschromatopsia that disease shows other characteristics that have been reported for congenital tritanopia. Dominantly inherited juvenile optic atrophy shows autosomal dominant inheritance with wide inter and intrafamilial variation of expression as has been claimed for congenital tritanopia, and the disease is claimed to have an incidence similar to that suggested by Wright for tritanopia. The finding by Grutzner that DIJOA is essentially a static disease is consistent with the nonprogressive nature of a congenital dyschromatopsia. On the other hand DIJOA is also characterized by temporal optic atrophy, reduced visual acuity, and visual field defects. Because these clinical symptoms and signs may be so mild and subtle as to go unnoticed by patient and examiner Krill, Smith, and Pokorny asserted that the presence of the disease has been missed in previous studies purporting to report congenital tritanopia.

In addition, they considered that the existence of a disease which serves as a model for congenital tritanopia when there are no diseases resembling congenital deutan and protan defects, requires a better explanation than mere coincidence. However, in our opinion there is an equally plausible alternative line of reasoning to suggest that congenital tritanopia should exist. We know there are three types of photopigment in human cones and Rushton has shown that two of these, chlorolabe and erythrolabe, are absent in deuteranopia and protanopia, respectively. Therefore, we might reasonably expect an inherited color vision anomaly arising from the congenital absence of cyanolabe to complete a trilogy of congenital dichromasies.

For this reason, and because we were not convinced by all of the arguments and evidence put forward by Krill, Smith, and Pokorny we decided to re-examine the (M) family of tritans reported by Henry, Cole, and Nathan and Cole, Henry, and Nathan in order to unequivocally confirm or exclude the presence of ocular disease. In addition, a second family of tritans which had been discovered by Henry, Cole, and Nathan was also investigated. Only one member of this (P) family has been previously reported; he was the subject RP in studies of mechanisms and color-naming in tritanopia.

Krill, Smith, and Pokorny laid down four conditions which they held must be met before the diagnosis of congenital tritanopia, as distinct from DIJOA, can be
made: (1) normal distance and near vision, (2) normal visual fields, (3) normal-appearing optic nerves in affected and unaffected family members, and (4) the ophthalmoscopic examination to be done by someone who has evaluated many pathologic optic nerves. (One might question whether they require too high a standard of examination for ocular disease when they are not prepared to accept the data of Schmidt; she was well aware of the need to exclude the presence of DIJOA, and the color-normal and tritan members of the family she reported underwent authoritative ophthalmologic examination.)

The above criteria can be extended: we chose to employ the eight clinical criteria proposed and confirmed by one of us as a sufficient and reliable diagnostic set quite specific for DIJOA. These criteria are: (1) dominant autosomal inheritance, (2) insidious onset in childhood, 4 to 8 years of age, (3) moderately reduced visual acuity, 6/20 to 6/60, (4) temporal pallor of the optic discs, (5) centrocecal enlargement of the blind spot, (6) full peripheral fields to white targets, (7) inverted peripheral fields to colored targets, and (8) acquired tritan dyschromatopsia.

These criteria are compared in Table I with those diagnostic characteristics expected of congenital tritanopia. Our analysis involved comparing the characteristics shown by four affected members of the M family and two affected members of the P family with the diagnostic criteria listed in Table I. If the tritans met all those criteria critical to the diagnosis of DIJOA then we could confirm that their dyschromatopsia was acquired secondary to that disease. If they met criteria which indicated the presence of ocular disease but were not specific for DIJOA we could confirm that their dyschromatopsia was acquired, but not necessarily secondary to DIJOA. If the only criteria met were those shown by Table I to be common to both DIJOA and congenital tritanopia then we could reasonably claim that their tritan defect was not acquired but is congenital. In addition three color-normal members of the P family were examined and information from case records of optometric examinations in 1968 and 1971 were available for a deceased member of the same family.

Method

A. The color vision of every subject was examined, each eye separately, using the Nagel anomaloscope, the Ishihara, AOHRR, and F2-pseudoisochromatic plates, the Farnsworth D-15 and 28 hue tests, and the Farnsworth Munsell 100 hue test. The nonspectral tests were administered following standard procedures using a Macbeth Easel lamp.

Acquired dyschromatopsias fluctuate in severity with progression or remission of their causative pathology whereas congenital defects are stable. Therefore, any alteration of a dyschromatopsia with time indicates an acquired origin, a useful criterion for differentiation when congenital onset is so difficult to establish. Admittedly, with the present patients, stability of dyschromatopsia would not necessitate congenital origin because the most likely pathologic cause, DIJOA, is an essentially nonprogressive condition. However, if an active disease process was involved progression could have produced dichromasy in a previously trichromatic patient, or remission produced trichromasy in a previously dichromatic patient; if such change was found, their dyschromatopsia can only be of acquired origin. We explored this possibility by repeating the test of dichromasy which had been administered to four of the six tritans in the course of earlier investigations. The test involves matching at the Donaldson filter colorimeter a mixture of red (dom. X 650 nm.) and blue (dom. X 452 nm.) to green (X 530 nm.): the dichromatic tritan can find a mixture of red and blue which appears identical to the green because the green and the mixture point lie on the same tritanopic confusion line. Normal observers and trichromatic tritans cannot make a match.

B. Visual acuity was measured on an American Optical Company projection chart with a background luminance of 88 cd. M−2 and letter-background contrast of 89 per cent. An externally illuminated Snellen chart was taken to the home of some of the color-normal subjects, and the color-normal child was examined with an illiterate E chart.

The visual acuities of two tritans were also measured on standardized psychometric visual acuity targets identical to the "S" charts used by Flom, Weymouth, and Kahneman. We used the apparatus of a colleague who was investigating visual acuity and details of the standardized test procedure will be found in his report.
Table I. Diagnostic characteristics of congenital tritanopia and tritanopia acquired secondary to dominantly inherited juvenile optic atrophy

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Congenital tritanopia</th>
<th>Tritanopia secondary to DIJOA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Color vision defect—tritanopia</td>
<td>Loss of color discrimination along chromaticity loci converging 42 to a point ( x = 0.171, y = 0.000 ) on the 1931 CIE chromaticity diagram such that the neutral point to illum. B is 571.5 nm, and 450 nm. matches 530 nm. Some congenital tritans show trichromatic vision and have been called incomplete tritanopes.</td>
<td>Dichromatic form simulates congenital tritan dichromasy, but is not always attained because the defect may have trichromatic or monochromatic stages. All stages are associated with red-green color confusions and disturbance of red-green metameric matches.</td>
</tr>
<tr>
<td>Inheritance</td>
<td>Autosomal dominant with partial expression common.</td>
<td>Autosomal dominant with variable expression usual</td>
</tr>
<tr>
<td>Onset</td>
<td>Congenital</td>
<td>Juvenile, insidiously at 4 to 8 years of age</td>
</tr>
<tr>
<td>Visual acuity</td>
<td>Normal</td>
<td>Moderately reduced, 6/20 to 6/60</td>
</tr>
<tr>
<td>Optic discs</td>
<td>Normal</td>
<td>Temporal optic atrophy</td>
</tr>
<tr>
<td>Visual fields</td>
<td>Central: Normal; Peripheral: White colors</td>
<td>Centrocecal enlargement of blind spot</td>
</tr>
<tr>
<td></td>
<td>Constricted to blue</td>
<td>Constricted to blue and less so to green</td>
</tr>
</tbody>
</table>

Table II. Clinical characteristics in patients from two pedigrees showing familial tritanopia

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (years)</th>
<th>Color vision</th>
<th>Visual acuity</th>
<th>Optic discs</th>
<th>Visual fields</th>
</tr>
</thead>
<tbody>
<tr>
<td>M-III-8</td>
<td>50</td>
<td>Tritan</td>
<td>6/4.5</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>M-III-11</td>
<td>40</td>
<td>Tritan</td>
<td>6/4.5</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>M-IV-6</td>
<td>25</td>
<td>Tritan</td>
<td>6/4.5</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>M-IV-7</td>
<td>12</td>
<td>Tritan</td>
<td>6/4.5</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>P-II-4</td>
<td>57</td>
<td>N</td>
<td>6/6</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>P-II-3*</td>
<td>555</td>
<td>Not known</td>
<td>6/4.5</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>P-III-2</td>
<td>27</td>
<td>Tritan</td>
<td>6/4.5</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>P-III-3</td>
<td>25</td>
<td>N</td>
<td>6/4.5</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>P-III-4</td>
<td>23</td>
<td>Tritan</td>
<td>6/4.5</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>P-IV-1</td>
<td>4½</td>
<td>N</td>
<td>6/6</td>
<td>N</td>
<td>N</td>
</tr>
</tbody>
</table>

There was no difference between the two eyes of each patient and therefore the entries in the Table refer to both the right and left eyes of each patient: tri, trichromatic; di, dichromatic; N, normal; con, constricted; +, deceased.

*Data from case records 1968 and 1971.

Cole and Watkins (1967).


gets consist of randomly oriented Landolt-C's within a framework of illiterate E's, and while such targets produce complex but constant contour interaction, they eliminate the influence of variation of contour interaction between adjacent Snellen letters on visual acuity. Amblyopic eyes perform relatively poorly compared with normal eyes on such targets. Background luminance of the chart was 91 cd. m.<sup>-2</sup> and letter-background contrast was 85 per cent. Comparison of performance by the two tritans on the AO projection Snellen chart and the psychometric charts gave us an effective calibration of the former. C. The optic discs and surrounding posterior and equatorial regions of the fundi were examined independently by each of the authors. One of us is an ophthalmologist; another of us has had first-hand experience of the fundus appearance in three families affected with DIJOA, and the other has observed several members from those families.

D. Central visual fields were measured on a 2 m. blackcloth Bjerrum screen with 2 mm. white and 5 mm. blue and red targets. The screen had an average luminance of 0.44 cd. m.<sup>-2</sup>. E. Peripheral fields were measured on a 0.3
Results

Results are summarized in Table II.

1. Inheritance. The genealogic trees are presented in Fig. 1. The M family is as reported in 1964\textsuperscript{11} and 1966\textsuperscript{12} except that M-IV-7 who was an infant at that time was found by us to have a tritan defect. Subject P-III-4 is RP of two previous studies\textsuperscript{22,23} but his tritan sister, P-III-2, has not previously been reported.

Inheritance of the tritan defect in the M family is undoubtedly dominant autosomal and the distribution of the tritan defect in the P family is consistent with the same form of inheritance. The mother of the tritans, the deceased P-II-3, was presumably the affected or carrier member of the second generation.

Phenotypic variation in the expression of the color-vision defect was present in both families.

2. Onset. None of the tritans recalled an acute onset of their color vision defect but most did recall being mildly aware of color confusions during early school years. On the other hand, neither twelve-year-old M-IV-7 nor his immediate family were aware of his color vision defect. Walls\textsuperscript{10} has pointed out that little credence can be given to patient reminiscence after they have been informed that their color vision is defective.

3. Color vision. The tritan defect described for M-III-8, M-III-11, M-IV-6, and P-III-4 in earlier studies\textsuperscript{11-12} was confirmed, and a tritan defect was detected in M-IV-7 and P-III-2.

Two significant points emerge from the results of the color vision examination. First, there was no change in the state of trito- or dichromasy of those patients who were so assessed in 1966\textsuperscript{11} and 1967\textsuperscript{22} (Table II). That is, Patients M-III-11 and P-III-4 are still dichromatic and M-III-8 and M-IV-6 have remained trichromatic. Patient P-III-2 fleetingly achieved the dichromatic match but the match was unstable. Her performance resembled that seen in extreme anomalous trichromasy and she is best described as an incomplete tritanope. Secondly, the clinical tests of color vision elicited no response which suggested there...
was secondary involvement of red-green perception. Not one single plate of the Ishihara tables nor the red-green series of the AOHRR was failed by any of the tritans. Settings on the Nagel Anomaloscope were always within normal limits and the matching range was narrow.

Unfortunately there is no written record of the color vision of the deceased P-II-3. Anamnesis of the family suggests her color vision was normal: they remember she gave normal responses to a battery of color vision tests (probably including the Panel D-15 and F2 plate) administered to the family by G. H. Henry when P-III-4 first came to notice.

The effectiveness of the individual tests in detecting and describing tritanopia varied, and is worth noting.

The Farnsworth F2 plate was failed by all six tritans, five of them seeing only the blue square and the sixth, M-IV-7, seeing both squares but the blue square as very much brighter. The only plate of the AOHRR on which failure occurred was the second blue-yellow screening plate, five of the twelve tritan eyes missing the triangle. The four diagnostic blue-yellow plates were always passed, but every subject reported that the tritan symbols on those plates were very much fainter than the corresponding tetartan symbols.

The two tritanopes made numerous clear-cut diametrical errors of the tritan type on the Panel D-15 and Farnsworth Munsell 28 hue tests. On those tests, the tritanomal's performance varied from a single transposition or several nondiagnostic transpositions to diametrical tritan errors fewer in number than that made by the dichromats. An unequivocal tritan axis was evident on plots of the Farnsworth Munsell 100 hue test error scores for all patients.

4. Fundi and optic discs. The fundi and optic discs of the tritan and color-normal family members appeared normal. A familial characteristic in both families was slight asymmetry of optic cup size but there was no temporal pallor of abnormal appearance. All three examiners agreed without difficulty. There was no appearance of abnormal macular pigmentation or degenerative or inflammatory macular change.

5. Visual acuity. The monocular visual acuity of the tritan and color-normal family members was in every case 6/6 or better. In the experiment with psychometric targets, both M-III-8 and M-IV-7, at the 50 per cent correct level, achieved a Snellen equivalent of better than 6/5. In the case records of P-II-3 visual acuities of 6/4.5 o.u. were recorded in October, 1971, when she was 55-years-of-age.

6. Visual fields. Visual fields were measured in five tritans; central and peripheral fields were in every case intact to white targets. In the history of P-II-3 it is noted that her central fields were normal to 2/2,000 mm. white targets in March, 1968. The blue chromatic field of the tritans was severely constricted, more so temporally, to lie within normal red and green isopters. The blue achromatic field generally coincided with the green achromatic isopter or lay between it and the red.

The five tritans extensively examined clearly met only four of the eight criteria diagnostic of DIJOA (inheritance, tritan dyschromatopsia, full white, and constricted blue field), but the criteria met are those which that disease has in common with congenital tritanopia (Table I). A blue-yellow color defect was the only functional loss found, and there was no evidence of optic atrophy, in particular nothing to suggest a diagnosis of DIJOA. There was no evidence of any macular or other ocular disease.

These results show no evidence that the tritan defect of color vision in the present patients was acquired, and we are directed to the conclusion that inherited congenital tritanopia does exist.

Discussion

When Krill, Smith, and Pokorny14, 15 questioned the existence of congenital tritanopia they did nothing new, but merely drew together a collection of long known facts to restate a once commonly held belief. The existence of tritan confusion lines

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and a convergence locus in acquired tritanopia has been known since König's study in 1897. In 1935, Riedl demonstrated that the color impairment in DIJOA primarily affects blue perception, and Jaeger in 1956 pointed out that the acquired tritan defect in DIJOA reaches dichromasy ("a dichromatic system with a neutral point in the yellow"). Thus, if the acquired dyschromatopsia of DIJOA is tritan, then it is not surprising that it can reach a dichromatic stage akin to congenital tritanopia, although it appears to have surprised Krill, Smith, and Pokorny who remarked "the major point is that when the neutral points and color matches of our subjects were plotted on the chromaticity diagram, convergence to the blue end of the spectrum in the area of the tritanopic-convergence locus was found." It is not meaningful to argue for the nosologic or etiologic sameness of congenital tritanopia and DIJOA using as evidence the "identical" color-vision defects. Many other diseases would qualify by this argument because whenever an acquired tritan defect occurs it has the potential to simulate congenital tritanopia. Indeed this point is well made by Cox's study of 9 acquired tritans whose defects were secondary to macular degeneration or retinopathy. She concluded that their "acquired condition has many points identical with congenital tritanopia."

Clearly the only justification for the assertion that congenital tritanopia has been confused with the acquired tritanopia of DIJOA is the subtleness with which that disease can occur and the possibility that it might previously have been overlooked. We certainly agree that this might have occurred in some of the earlier reports of tritanopia and there is one likely example: in his study of congenital tritanopia Wright presented as a "special case of color defect" a tritan observer whose hue discrimination operated only at the red end of the spectrum. Jaeger and Grutzner have shown that such hue discrimination can occur in the tritan defect secondary to DIJOA. Wright gave no clinical data on this subject but it does appear likely that he was affected with DIJOA. If this were so then the acquired and congenital forms of tritanopia were not really confused, because Wright found it necessary to distinguish the character of the acquired form from that of congenital tritanopia.

The extensive and sophisticated laboratory studies reported by Krill, Smith, and Pokorny in their 1971 paper provide solid evidence of the extent to which acquired tritanopia may sometimes mimic congenital tritanopia. On the other hand, their data on AOHRR performance and neutral point size illustrate two basic differences between the two forms.

Several of their acquired tritans made substantial failures on those plates of the AOHRR designed for the detection of red-green defects. However, not one of the six congenital tritans in our study failed a single pseudoisochromatic plate intended for red-green defects, and no other reported congenital tritans have performed any differently from color-normal subjects on such plates. In fact Judd, Plaza, and Farnsworth said "... performance on all tests for the detection of red-green blindness was above average." The data of Krill, Smith, and Pokorny substantiates previous findings of common failure on red-green tests of the AOHRR by patients with the tritanopia of DIJOA. Smith has previously pointed out that for this reason the AOHRR is a misleading test for the diagnosis of the tritan defect in DIJOA: its use caused Caldwell, Howard, and Riggs to report "both blue-yellow and red-green defects were present;" and Kok-van Alphen was led to report that the defect in some cases was of red-green and not of blue. Krill, Smith, and Pokorny acknowledged the presence of concomitant red-green confusions but did not recognize their significance; they stated "in pedigrees with a tritanope there are frequently some affected individuals showing evidence of slight red-green abnormalities on the pseudoisochromatic plates and 100 hue test" and cited the Henry, Cole, and Nathan.
pedigree. However, examination of the latter reveals that the red-green abnormality they refer to was a typical deuteranope who presumably inherited his defect in the usual sex-linked way from his grandfather who had married into an unaffected branch of the family.

The second point of difference is the character of the neutral point. Congenital dichromats are reported to make the spectral setting corresponding to white very accurately such that it is very close to a point on the spectrum. Acquired tritanopes have a range of spectral settings which correspond to white, that is they have a neutral zone. Krill, Smith, and Pokorny measured neutral points in 11 eyes: in one of these it was a point, in the other 10 it was a zone between 3 nm. and 13 nm. wide, with an average width of 5.5 nm. Thus, in another respect the dyschromatopsias in question are not identical.

Congenital tritanopes are also different from the acquired tritans of DIJOA in the way they color-name spectral colors. In an investigation of the color naming of M-III-8, M-IV-6, and P-III-4, one of us showed that they confuse blue and green such that the name "blue" can replace the name "green" in the green portion of the spectrum. That they do not see the spectrum as desaturated is indicated by their minimal use of the color name "white." In an unpublished study of color-naming in DIJOA the results of Fig. 2 were obtained. The technique used was identical to that reported in a study of color-naming by normal subjects except for the allowance of the extra response name "white." Subjects were H-III-4 and H-III-6 of the previously reported pedigree of DIJOA. There is no confusion between blue and green and the individual color names are employed at the spectral loci appropriate to the normal response. However, the amplitude of the color name responses is depressed because of severe desaturation of the spectrum at short and middle wave lengths. The character of their color-naming is distinctively different from that in congenital tritanopia and not readily accountable for by differences in experimental method; they do not confuse blue and green color names as do congenital tritans, but unlike congenital tritans they do see the shorter wave lengths as desaturated. This briefly described difference in color-naming behavior is undoubtedly a reflection of the different physiologic mechanisms underlying the respective dyschromatopsias and additional evidence that the two conditions are not identical.

Several points arising from the present study deserve comment.

1. The mode of inheritance of tritanopia in the P family is not clear. P-I-2, the maternal grandfather of the tritans, is believed by the family to have had defective color vision. If this were so, and his daughter P-II-3 was unaffected as suggested by family anamnesis, then the tritanopia has skipped a generation and shows incomplete penetrance. On the other hand, if neither P-I-2 or P-II-3 were affected, then the tritanopia has been transmitted as an autosomal recessive, or else has arisen as a mutation into the third generation. The father of the tritans, P-II-4, has normal color vision.
and there is no history of dyschromatopsia in his family.

2. The presence of dichromatic and trichromatic forms of tritan defect in the M family has previously been noted and it appears that both forms are also present in the P family. This reinforces the conclusion reached by Cole, Henry, and Nathan that tritanomaly is a forme fruste of tritanopia, and for that reason is better termed incomplete tritanopia to distinguish its character from that of the red-green anomalous trichromacies which clearly have a separate mechanism and genetic basis from their dichromatic counterparts.

3. Some comment on the clinical tests of color vision is also appropriate. The AOHRR tables are a satisfactory test for congenital tritan defects only if the precaution is taken of asking the subject about the relative brightness of the symbols. None of the present tritans positively failed this test, but all did find the tritan symbols considerably fainter than the tetertan symbols on the same plate. This affirms the previous experience of Cole. The Farnsworth F2 plate, which can be easily made up from Munsell papers according to the description given by Kalmus was positively diagnostic for all tritans, and more effective than all 6 plates of the AOHRR. In conjunction with an efficient screening test like the F2 plate, Panel D-15 performance may be a useful index of severity. In the same way that red-green defectives are practically differentiated by the D-15 according to the severity of their defect, both tritanopes failed the D-15, whereas the incomplete tritanopes performed well or almost normally.

The significance of the settings on the Nagel Anomaloscope by patients with DJIOA is not clear. The patients of Krill, Smith, and Pokorny made virtually normal settings, while others have found deutan and protan settings. These findings may not be incompatible: while the primary loss is of the mechanism subserving blue perception, concomitant impairment of red or green perception will occur if the abiotrophic process extends to involve the red or green mechanisms, and which of these is greater affected may determine the direction of the secondary confusion axis.

We conclude that inherited congenital tritanopia does exist, and suggest that it will be found more commonly if clinicians use suitable tests such as the F2 plate and the D-15 for its detection.

REFERENCES


