Retinal vein occlusion (RVO) is a common cause of visual impairment. The Eye Disease Case-Control Study identified hypertension, diabetes mellitus, cardiovascular disease, and glaucoma as risk factors for RVO. However, vein occlusion before age 40 is relatively uncommon. Most studies show that only 10% of all cases of RVO occur in patients younger than 40 years of age. Because of this, conclusions from larger studies such as the Eye Disease Case-Control Study may not be applicable to this younger population.

In addition, the relatively few reports of RVO in younger patients have sometimes offered contradictory conclusions. For example, with respect to visual outcome, in a study of 17 patients younger than age 40 years with central RVO (CRVO), Walters and Spalton concluded that visual prognosis in this population was good; only one patient (5.8%) lost vision. The authors found little evidence of underlying vascular disease in most patients, although 10 patients (59%) had thrombotic risk factors.

In contrast, more recently, Recchia and colleagues, looking at a slightly older and larger cohort (57 patients 55 years of age or younger), concluded that visual prognosis in CRVO is variable. Of 22 patients presenting with a visual acuity (VA) of 20/40 or better, eight (36%) declined to 20/400 or worse during the course of follow-up. Conversely, of 10 patients presenting with a VA of 20/200 to 20/400, eight (80%) improved to 20/60 or better. Overall, 28% of patients lost vision during follow-up. In this study, 40 patients (70%) had associated diseases.

Similarly contradictory findings can be seen in laboratory evaluations of patients with RVO. For example, Gottlieb and colleagues found no increase in the

**Figure 1. A plot of initial VA against final VA in all patients in the study.**
prevalence of resistance to activated protein C or factor V Leiden in a study of blood samples from CRVO patients younger than 50 years. Their finding of APC resistance in only one of 21 patients (4.7%) was similar to that found in a general population.

In contrast, Lahey and colleagues found that the percentage of positive hypercoagulable tests was higher in a study population with CRVO than age-matched controls. Fifteen of 55 patients (27%) younger than 56 years of age had one positive test result suggesting hypercoagulability. They found that hyperhomocysteinemia and circulating antiphospholipid antibodies were significantly more common in CRVO patients than in age-matched controls.

**STUDY DESIGN**

To further examine these issues, and to contribute to the understanding of RVO in young patients, we performed a study to assess visual outcomes in eyes with RVO in patients age 40 years and younger, and to identify possible association with systemic risk factors. This retrospective, consecutive case series included patients diagnosed with branch RVO (BRVO), hemiretinal vein occlusion (HRVO), or CRVO presenting to a single retina practice over 11 years, from January 1997 to December 2007. Patients 40 years of age and younger with more than 1 month of follow-up were included, and only one eye per patient was enrolled.

We enrolled a total of 39 eyes of 39 patients, 62% of whom were male, and whose mean age was 32 years. Mean duration of symptoms was 33 days, and mean length of follow-up was 20 months.

The diagnosis was CRVO in 27 eyes (69%), BRVO in eight eyes (21%), and HRVO in four eyes (10%). Thirty-two eyes (82%) were nonischemic at the time of presentation, and seven eyes (18%) were found to have evidence of retinal ischemia on fluorescein angiogram at presentation.

**RISK FACTORS**

Our results can be divided into three areas: risk factors identified, visual outcomes, and factors associated with visual prognosis.

Previously identified risk factors for RVO in older populations have included systemic conditions such as hypertension, hyperlipidemia, diabetes mellitus, obesity, and use of oral contraception. In the present study population, 29 patients (74%) had at least one of these risk factors, and 11 patients (28%) had multiple risk factors. Ten patients (26%) had no identifiable risk factors. In addition, nine patients (23%) had an abnormal hypercoagulable workup.

The most common risk factors seen in this population were hypertension in 11 patients (28%), use of oral contraception medication in six (15%), and hyperlipidemia in four (10%). Other risk factors seen in less than 10% of patients included diabetes, obesity, smoking, and glaucoma.

The most common laboratory abnormality identified was a mutation in the methylenetetrahydrofolate reductase (MTHFR) gene in four patients (10%). The product of this gene encodes a protein involved in homocysteine metabolism. Three patients (8%) were also found to have mutations in the prothrombin gene.

**VISUAL OUTCOMES**

Most eyes in this study had a good visual outcome (Table 1). The mean baseline VA was 20/50. Mean VA at

<table>
<thead>
<tr>
<th>TABLE 1. VISUAL OUTCOMES</th>
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<tbody>
<tr>
<td><strong>Baseline</strong></td>
</tr>
<tr>
<td>Mean VA</td>
</tr>
<tr>
<td>Range</td>
</tr>
<tr>
<td>Improved VA</td>
</tr>
<tr>
<td>Stable VA</td>
</tr>
<tr>
<td>Loss of VA</td>
</tr>
</tbody>
</table>

VA = visual acuity
LP = light perception
NLP = no light perception
the final visit was 20/40. At the final visit, 20 eyes (51%) had improved VA, 10 (26%) had stable vision, and nine (23%) lost vision.

Figure 1 shows a plot of initial VA against final VA in all patients in the study. Most of the data points are in the area above the line, indicating visual improvement. In addition, more than 80% of patients had a final VA of 20/60 or better—those to the right of the red line.

To identify factors predictive of VA outcome, we compared the characteristics of patients whose eyes had a final VA of 20/60 or better with those of patients whose eyes had worse than 20/60 final VA. This analysis showed that patients whose eyes had a final VA worse than 20/60 were more likely to have worse presenting baseline vision and more likely to have evidence of retinal ischemia on fluorescein angiography (Table 2). In contrast, no significant differences were seen between the two groups with respect to duration of symptoms, age, or number of risk factors.

**CONCLUSIONS**

This study was limited by its small size and nonrandomized design. In addition, it lacked a control group, and measurement of VA was not standardized.

We found that most patients with RVO (82%) presented with good VA (mean 20/50) and nonischemic disease. In addition, most patients (77%) improved from or maintained their baseline vision during follow-up that ranged from 1 month to more than 10 years, and most patients obtained a good final VA (mean 20/40).

Poor baseline VA and retinal ischemia were factors associated with worse VA outcome. There was a strong association between RVO and systemic risk factors (74%) in these young patients, and almost one-fourth (23%) had an abnormal hypercoagulable workup.

Based on the study findings, the authors recommend that young patients presenting with RVO should undergo extensive evaluation to identify possible systemic risk factors for thrombosis, including the use of prothrombotic medications and undiagnosed hypercoagulable status. Further, initial evaluation in these patients should emphasize baseline VA and retinal perfusion status, as these are strong predictors of final visual outcomes.

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**TABLE 2. FACTORS ASSOCIATED WITH VISUAL PROGNOSIS**

<table>
<thead>
<tr>
<th></th>
<th>20/60 or better</th>
<th>Worse than 20/60</th>
<th>Probability value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baseline VA (mean)</td>
<td>20/30</td>
<td>20/200</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Retinal ischemia on FA</td>
<td>6%</td>
<td>62%</td>
<td>.0017</td>
</tr>
<tr>
<td>Symptom duration (mean months)</td>
<td>32</td>
<td>32</td>
<td>.42</td>
</tr>
<tr>
<td>Age (mean years)</td>
<td>37</td>
<td>17</td>
<td>.89</td>
</tr>
<tr>
<td>Number of risk factors (mean)</td>
<td>1</td>
<td>1</td>
<td>.89</td>
</tr>
</tbody>
</table>

VA = visual acuity
FA = fluorescein angiography