

onstrated a bilateral temporal retinal disturbance characterized by increased inner-retinal reflectivity without a significant increase in retinal thickness (Figure 2). Multifocal electroretinography (mfERG) revealed evidence of bitemporal retinal cone-mediated pathway dysfunction (Figure 2), whereas full-field electroretinography and visual evoked responses were normal in both eyes. Magnetic resonance imaging and magnetic resonance angiography were normal.

Over the next six weeks, although the patient's complaints of binasal field defects remained unchanged, funduscopy demonstrated narrowing and whitening of the arterioles in the temporal macula of both eyes associated with a few, small intraretinal hemorrhages, without retinal thickening or edema. Repeat fluorescein angiography continued to demonstrate normal foveal avascular zones in both fundi, although in the far temporal macula, there were large areas of arteriolar occlusion with capillary dropout (Figure 3). Repeat OCT demonstrated bilateral temporal retinal thinning in the parafoveal retina without visible retinal changes on ophthalmoscopy or angiography (Figure 3).

Because our patient presented seven days after onset of symptoms, we did not recommend any specific therapy and instead advised follow-up with a hematologist regarding potential prophylactic systemic therapy.

Macular occlusive disease occurs more commonly in SS disease than SC disease<sup>1-4</sup>; however, our patient developed bilateral macular ischemic and frank retinal arteriolar occlusive disease in the setting of SC disease. Equally of interest is that the manifestation of this patient's retinal dysfunction was that of bilateral nasal field defects that, in the presence of a normal retinal examination and normal fluorescein angiography, raised the suspicion of bilateral optic nerve dysfunction.

Although retinal lesions uncommonly cause binasal field defects, the OCT in our patient revealed early retinal abnormalities and the mfERG confirmed a bitemporal retinopathy. Although the patient had symmetrically large optic nerve cups, in the setting of low intraocular pressures and healthy appearing peripapillary nerve fiber layer, the cupping is believed to be physiologic, and the sudden onset and spacial pattern of the visual field defects are consistent with the retinal lesions noted on OCT and mfERG. This case demonstrates that OCT and mfERG may be useful adjuncts in distinguishing optic nerve dysfunction from early ischemic retinal disease in the absence of ophthalmoscopic or fluorescein angiographic evidence of such disease.

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## Diagnostic Ability of Optical Coherence Tomography with a Normative Database to Detect Band Atrophy of the Optic Nerve

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**PURPOSE:** To evaluate the diagnostic ability of stratus optical coherence tomography (OCT) with a normative database to detect band atrophy (BA) of the optic nerve. **DESIGN:** Cross-sectional study.

**METHODS:** StratusOCT retinal nerve fiber layer thickness scans were obtained in 37 eyes with BA and permanent temporal visual field defect and 37 healthy eyes. The categorization of eyes according to the normative database of the instrument and sensitivity and specificity values were reported.

**RESULTS:** The average thickness parameter demonstrated the highest sensitivity for detection of abnormalities in eyes with BA, followed by the parameters related to the temporal and nasal quadrants. Values of sensitivity were relatively lower for the 30 degree segments.

**CONCLUSION:** StratusOCT with a normative database performed well in detecting retinal nerve fiber layer (RNFL) loss in eyes with BA. Clinicians should be aware of possible detection failure in the 30 degree segmental analysis, particularly at the 3 o'clock meridian. (*Am J Ophthalmol* 2007;143:896-899. © 2007 by Elsevier Inc. All rights reserved.)

**P**ATIENTS WITH CHIASMAL COMPRESSION AND TEMPORAL visual field (VF) loss often develop retinal nerve fiber layer (RNFL) loss in the nasal and temporal sectors of the optic nerve with relative preservation of

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**TABLE 1.** Relative Frequencies of Categorization in 37 Eyes With Band Atrophy of the Optic Nerve and 37 Normal Eyes, According to the Normative Database of the StratusOCT Optical Coherence Tomography

Parameter	Band Atrophy			Normal Eyes		
	Within Normal Limits	Borderline	Outside Normal Limits	Within Normal Limits	Borderline	Outside Normal Limits
Average thickness	1 (3%)	6 (16%)	30 (81%)	37 (100%)	0	0
Superior thickness	10 (27%)	4 (11%)	23 (62%)	37 (100%)	0	0
Temporal thickness	6 (16%)	6 (16%)	25 (68%)	37 (100%)	1 (3%)	0
Inferior thickness	9 (24%)	11 (30%)	17 (46%)	36 (97%)	1 (3%)	0
Nasal thickness	2 (5%)	21 (57%)	14 (38%)	35 (95%)	2/37	0
Thickness at 12 o'clock	22 (59%)	4 (11%)	11 (30%)	36 (97%)	1 (3%)	0
Thickness at 11 o'clock	20 (54%)	5 (14%)	12 (32%)	36 (97%)	1 (3%)	0
Thickness at 10 o'clock	10 (27%)	6 (16%)	21 (57%)	37 (100%)	0	0
Thickness at 9 o'clock	6 (16%)	16 (43%)	15 (41%)	36 (97%)	1 (3%)	0
Thickness at 8 o'clock	10 (27%)	7 (19%)	20 (54%)	36 (97%)	1 (3%)	0
Thickness at 7 o'clock	26 (70%)	2 (5%)	9 (24%)	37 (100%)	0	0
Thickness at 6 o'clock	27 (73%)	4 (11%)	6 (16%)	36 (97%)	1 (3%)	0
Thickness at 5 o'clock	7 (19%)	6 (16%)	24 (65%)	36 (97%)	0	1 (3%)
Thickness at 4 o'clock	8 (22%)	18 (49%)	11 (30%)	34 (92%)	3 (8%)	0
Thickness at 3 o'clock	20 (54%)	14 (38%)	3 (8%)	36 (97%)	1 (3%)	0
Thickness at 2 o'clock	7 (19%)	11 (30%)	19 (51%)	36 (97%)	1 (3%)	0
Thickness at 1 o'clock	6 (16%)	6 (16%)	25 (68%)	37 (100%)	0	0

the superior and inferior quadrants. This pattern of loss is called band atrophy (BA) of the optic nerve and is important in the diagnosis and management of chiasmal compression.<sup>1</sup> Recently, we and others have demonstrated that an earlier version of optical coherence tomography (OCT) is able to identify RNFL loss in eyes with BA.<sup>2,3</sup> The new StratusOCT (Carl-Zeiss Meditec, Dublin, California, USA) provides information on the probability of abnormality of patient examination results after comparison with an internal normative database. Although there has been considerable research on RNFL thickness measurements with this technology, there are few reports on the diagnostic ability of OCT with a normative database,<sup>4</sup> and none in eyes with BA. Because clinicians usually rely on the instrument's printout for identification of abnormalities, it is important to know whether the information conveyed by the printout accurately reflects the existence of RNFL loss in these patients. The purpose of this study was to evaluate the diagnostic ability of StratusOCT in eyes with BA using the comparison with its internal normative database.

Thirty-seven eyes from 37 patients with BA and 37 eyes from 37 age- and gender-matched healthy controls were studied. All patients with history of chiasmal lesions had already been submitted to previous treatment of the suprasellar lesion and had stable VF defects for at least one year before study entry. Inclusion criteria included visual acuity of 20/25 or better; spherical and cylinder refraction within  $\pm 4$  diopters (D), absence of glaucoma, reliable VFs, complete or partial temporal hemianopia on standard automated perimetry (SAP [Humphrey 24-2 SITA strategy]), and a normal nasal hemifield. Twenty-one eyes had

**TABLE 2.** Sensitivity and Specificity (with 95% CI) for StratusOCT Parameters, Based on the Internal Normative Database, in Eyes With Band Atrophy of the Optic Nerve and Normal Controls

Parameter	Sensitivity (95% CI)	Specificity (95% CI)
Average thickness	0.97 (0.92–0.97)	1.00 (0.94–1.00)
Superior thickness	0.73 (0.66–0.73)	1.00 (0.93–1.00)
Temporal thickness	0.84 (0.76–0.86)	0.97 (0.89–0.99)
Inferior thickness	0.76 (0.67–0.78)	0.97 (0.89–0.99)
Nasal thickness	0.95 (0.87–0.98)	0.95 (0.87–0.98)
Thickness at 12 o'clock	0.41 (0.32–0.43)	0.97 (0.89–0.99)
Thickness at 11 o'clock	0.46 (0.37–0.48)	0.97 (0.89–0.99)
Thickness at 10 o'clock	0.73 (0.66–0.73)	1.00 (0.93–1.00)
Thickness at 9 o'clock	0.84 (0.76–0.86)	0.97 (0.89–0.99)
Thickness at 8 o'clock	0.73 (0.65–0.75)	0.97 (0.89–0.99)
Thickness at 7 o'clock	0.30 (0.23–0.30)	1.00 (0.93–1.00)
Thickness at 6 o'clock	0.27 (0.19–0.29)	0.97 (0.90–0.99)
Thickness at 5 o'clock	0.81 (0.73–0.83)	0.97 (0.89–0.99)
Thickness at 4 o'clock	0.78 (0.69–0.83)	0.92 (0.82–0.97)
Thickness at 3 o'clock	0.46 (0.37–0.48)	0.97 (0.89–0.99)
Thickness at 2 o'clock	0.81 (0.73–0.83)	0.97 (0.89–0.99)
Thickness at 1 o'clock	0.84 (0.77–0.84)	1.00 (0.93–1.00)

CI = confidence intervals.

complete temporal hemianopia (mean SAP mean deviation [MD] =  $-4.09 \pm 1.59$  decibels [dB]) and 16 eyes had partial temporal hemianopia (mean SAP MD =  $-0.25 \pm 2.00$  dB). Approval from the local Institutional Review Board Ethics Committee was obtained.

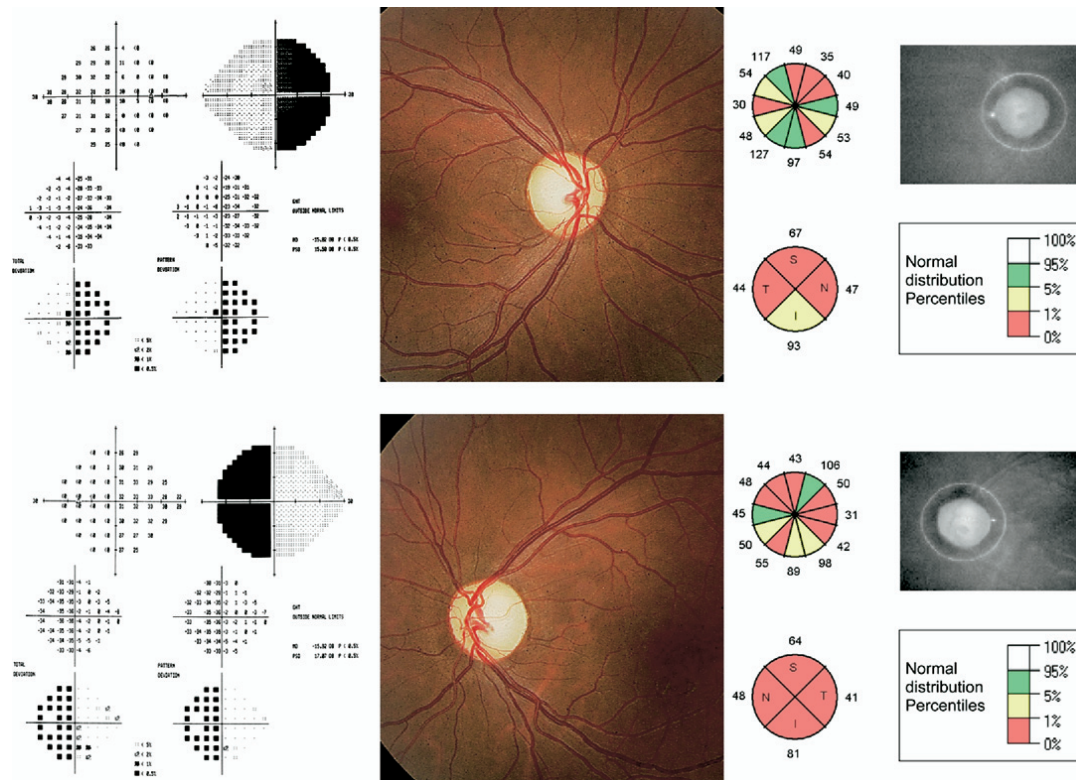


FIGURE. Visual field [VF] (Left), optic nerve photograph (Middle), and Stratus optical coherence tomography (OCT) results (Right) of a patient with band atrophy (BA) of the optic nerve (above: right eye; below; left eye). In each eye, the VF shows complete temporal hemianopia in agreement with the BA of the optic nerve. Note the retinal nerve fiber layer (RNFL) loss in the temporal and nasal portions of the optic nerve with relative preservation of the superior and inferior arcuate fibers. The StratusOCT printout shows the comparison with the normative database in each quadrant and in the 30 degree segments around the optic nerve. Loss of nerve fibers is indicated by outside normal limits results for several segments and quadrants. The 3 o'clock segments show a within normal limits result in both eyes.

Subjects underwent imaging with the StratusOCT, using the fast RNFL algorithm (software version 4.0.1). For each parameter, the StratusOCT software provides a classification (within normal limits, borderline, or outside normal limits) based on the comparison to an internal normative database of 328 eyes. A parameter is classified as outside normal limits if its value falls below the 99% confidence level of the normal, age-matched population. A borderline result indicates that the value is below the 95% confidence level, and a within normal limits result indicates that the value is higher than the lower 95% confidence level.

Table 1 shows the relative frequencies of categorization of eyes according to the normative database of the instrument. Values of sensitivity and specificity for the RNFL parameters are provided in Table 2, assuming borderline results as outside normal limits. The average thickness parameter demonstrated the highest sensitivity for detection of abnormalities in eyes with BA, followed by the parameters related to the temporal and nasal quadrants. Values of sensitivity were relatively lower for the 30 degree segments, especially for the ones located on the superior and inferior sectors of the optic disk, but also for the

thickness at 3 o'clock (nasal) (Figure). Specificity values were high for all RNFL parameters.

The low sensitivity found for the parameter thickness at 3 o'clock was particularly surprising. Because this segment presumably receives nerve fibers exclusively from the nasal retina, we would expect it to be abnormal in most BA patients. The lower performance of this parameter could be potentially explained by a larger variability in RNFL measurements at this segment,<sup>5</sup> making more difficult the detection of abnormalities.

In conclusion, StratusOCT parameters performed well in detecting RNFL loss in patients with BA when compared with the instrument's normative database. Sensitivity values, however, differed among the parameters and were higher for global- and quadrant-related measures than for 30 degree segmental analyses.

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## Rubella Virus–Associated Uveitis in a Nonvaccinated Child

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**PURPOSE:** To report presumed Fuchs heterochromic uveitis (FHU) associated with Rubella virus (RV)-specific intraocular antibody production in a child who was not vaccinated against rubella.

**DESIGN:** Observational case report.

**METHODS:** We examined a 13-year-old boy with chronic anterior uveitis complicated by mature cataract. Two aqueous humor (AH) samples taken with an interval of four weeks were analyzed for intraocular antibody production against RV by calculation of the Goldmann-Witmer coefficient.

**RESULTS:** The patient showed all the clinical signs for FHU: iris atrophy, stellate keratic precipitates, and cataract. Analysis of the AH demonstrated intraocular antibody production against RV in two sequential samples.

**CONCLUSIONS:** The data show that RV-associated uveitis can already present during childhood. Moreover, this finding suggests that nonvaccinated children may be at risk to develop uveitis after RV infection. (*Am J Ophthalmol* 2007;143:899–900. © 2007 by Elsevier Inc. All rights reserved.)

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WE DESCRIBE A CASE OF PRESUMED FUCHS HETEROCHROMIC uveitis (FHU) with rubella virus (RV)-specific antibody production in the aqueous humor (AH) in a nonvaccinated child.

A 13-year-old boy was referred because of unilateral uveitis complicated by mature cataract. Ophthalmologic examination revealed a visual acuity in the left eye of hand movements. Slit-lamp examination disclosed stellate keratic precipitates diffusely scattered across the corneal endothelium and mature cataract. Few cells were seen in the anterior chamber and one small posterior synechia was observed. No hyperemia of the conjunctiva was found. The patient was referred to a pediatrician, but there were no indications for systemic disease. Despite the small posterior synechia, the clinical characteristics suggested FHU. An AH sample taken for diagnostic purposes was negative for intraocular antibody production against Herpes simplex virus, Varicella zoster virus, and *Toxoplasma gondii*, but positive for RV, as determined by Goldmann-Witmer coefficient (GWC 31.93). The parents indicated that the patient had been refrained from vaccination for religious reasons, but they did not recall their child having had rubella. One month later, the patient's cataract was treated by phacoemulsification and intraocular lens implantation. The optimal postoperative visual acuity of the left eye was 6/5 (Snellen). A second AH sample collected during surgery was also positive for RV-specific antibody production (GWC 114.23).

The absence of posterior synechiae before surgery is considered to be one of the criteria of FHU.<sup>1</sup> However, because the incidence of pediatric FHU is low (reportedly less than 1%),<sup>2</sup> little is known about the clinical presentation of FHU in this age group. Possibly, posterior synechiae are formed in RV-induced FHU in the pediatric population.

RV infection may occur without clinical signs; the incidence of subclinical infections has been estimated as high as 25%.<sup>3</sup> The most important complication is a prime infection during pregnancy, which may give rise to congenital rubella syndrome (CRS) in the unborn child. In addition to severe systemic consequences such as cardiovascular defects and deafness, CRS may also include serious ocular abnormalities such as congenital cataract and pigmentary retinopathy.<sup>3,4</sup>

With the introduction of vaccination against RV in The Netherlands (since 1974 for school children and 1987 for all 14-month-old children), the incidence of rubella and CRS has declined significantly.<sup>4</sup> However, because of the presence of antivaccination groups based on religious incentives, rubella epidemics still occur in The Netherlands.<sup>4</sup>

The relationship between FHU and RV is not yet clear. Intraocular antibody production against RV is seen in almost 100% of FHU cases.<sup>5</sup> This might suggest that the intraocular immune response against RV is involved in the pathogenesis of this type of uveitis. One may speculate that