experienced trainer must use his or her discretion for a few complicated cataracts, such as those involving zonular dehiscence or microphthalmos. It also would be unlikely that a trainee operate on such complex cases. In this rubric, each step is individually assessed and graded. It also encourages teaching and assessment following good principles of feedback. Further work will be necessary to validate videotape surgeries.

The ICO has approved this assessment tool (e-Supplement 1, available at jaapos.org) The specific behavioral narrative anchors in the rubric provide raters with objective benchmarks for comparative purposes and provides learners with specific targets for behavioral change.

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Autoinflammatory retinopathy in chronic infantile neurological cutaneous and articular (CINCA) syndrome

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Chronic infantile neurological cutaneous and articular (CINCA) syndrome is a rare autosomal dominant autoinflammatory disease. We report the cases of monozygotic twins with CINCA syndrome whose predominant ocular manifestation was inflammatory rod-cone retinal dystrophy. Atypically, there were significant differences between twins in phenotype severity, suggestive of epigenetic differences and/or involvement of environmental factors.

Case Reports

Male twins, born at 38 weeks' gestation with normal birth weights and having monozygosity confirmed by genetic testing, were diagnosed by age 6 months with familial Mediterranean fever after suffering from ongoing fevers, recurrent chronic urticarial rashes, pericarditis, and recurrent bilateral destructive large joint inflammation and effusions. The phenotype for twin 1 was generally more severe than for twin 2, with progression of global developmental delay, chronic aseptic meningitis, and sensorineural hearing loss. The mother had observed intermittent episodes of bilateral, painless, red eyes without visual disturbance or photophobia in both twins from 5 years of age. All episodes resolved spontaneously; however, due to the recurring nature of the condition (2-3 weeks approximately every 4 months), they were referred for ophthalmology examination.

The twins presented to Brisbane Mater Children's Hospital for their first evaluation at 9 years of age. On examination, twin 1 had best-corrected visual acuity of 6/6 in each eye, with no relative afferent pupillary defect and normal color vision (Ishihara plates). Dilated fundus examination revealed mildly swollen optic nerve heads, thought to be related to aseptic meningitis.

Twin 2 had best-corrected visual acuity of 6/6 in each eye. The recurrent episodes of red eyes were diagnosed as recurrent episcleritis. Nine months later, twin 1 experienced one episode of mild anterior uveitis, successfully treated with a 6-week tapering course of topical fluorometholone.

When they were 11 years old, genetic testing confirmed a mutation in the *NLRP3* gene (T348M subtype), and the diagnosis was revised to chronic infantile neurological cutaneous and articular (CINCA) syndrome. For both twins, immunosuppressive treatments (oral corticosteroid and colchicine) were replaced with biweekly subcutaneous administration of anakinra. The response to treatment was excellent for all rheumatological aspects, with complete resolution of recurrent episcleritis in both twins.

When they presented for follow-up examination at age 14 years, their mother noted that twin 1 was become increasingly clumsy and was frequently bumping into objects. Direct questioning revealed that twin 1 also had symptoms of worsening nyctalopia. On examination, twin 1's best-corrected visual acuity was 6/9 in each eye. Both pupils reacted sluggishly. Color vision remained normal; anterior segments were quiet. The retina appeared atrophic, with attenuated arterioles. There was marked pigmentation of the macular surrounded by a ring of depigmentation in the parafoveal region (Figure 1). No retinal pigment epithelial migration was seen in the periphery.

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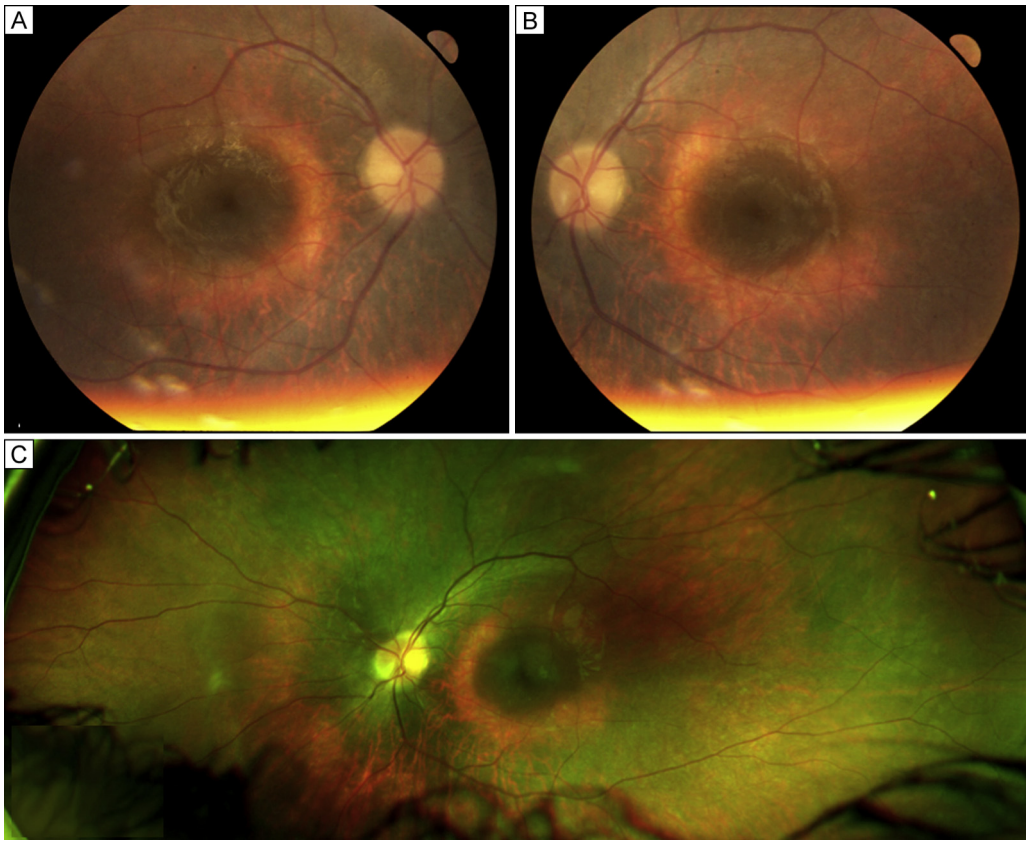

FIG 1. Fundus photographs of twin 1 at age 13 showing atrophic retina with attenuated arterioles and paramacular ring of chorioretinal atrophy. A, Right eye. B, Left eye. C, Wide-angle Optos retinal image of the left eye of twin 1.

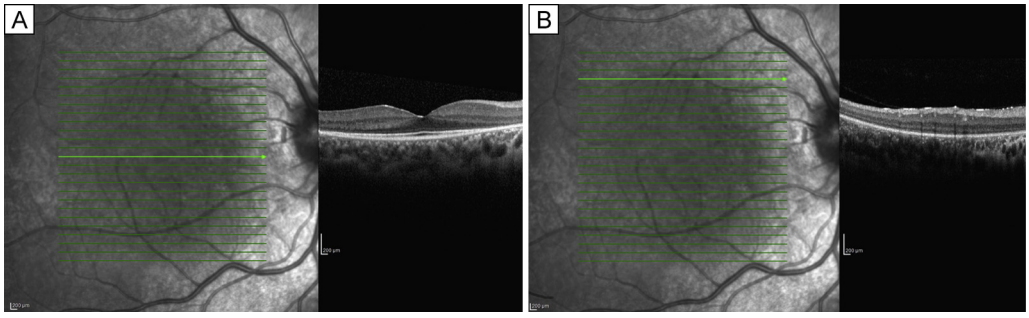

FIG 2. SD-OCT showing parafoveal thinning of the outer nuclear layer and loss of the ellipsoid layer in the right eye of twin 2. A, Foveal/parafoveal slice. B, Parafoveal slice.

The Humphrey 30-2 visual field showed advanced peripheral constriction in both eyes.

Visual acuity and clinical examination in twin 2 were normal; however, visual fields showed peripheral visual field loss in both eyes. Fundus autofluorescence showed parafoveal hyperfluorescent rings in both twins, more notable in twin 2. Spectral domain optical coherence tomography (Spectralis; Heidelberg Engineering, Heidelberg, Germany) showed thinning of the outer nuclear layer and loss of the ellipsoid layer in the parafoveal region, corresponding to the hyperfluorescent rings on autofluorescence (Figure 2).

Full-field (RETI-port gamma, Roland Consult, Brandenburg Germany) and multifocal electroretinography (ERG; RETI-port gamma) were performed according to International Society for Clinical Electrophysiology of Vision 2014 standards. In twin 1 testing demonstrated minimal residual function with the single flash (both