Diagnosis	Comment	Lab Testing
Behcet's disease	Typically a bilateral, recurrent, potentially blinding panuveitis associated with retinal vasculitis	No definitive test; strictly a clinical diagnosis
Birdshot retino-choroidopathy	A chronic, bilateral intermediate and posterior uveitis very strongly associated with HLA A29 and frequently treated by chronic immunosuppression	Almost all patients are HLA A29+
Blau syndrome	A very rare, autosomal dominant form of uveitis associated with mutations in the nucleotide binding domain of *NOD2*, a gene which codes for an important component of the innate immune system which recognizes bacterial cell wall	Genotyping identifies the mutations known to be associated with this syndrome
HLA B27-related:	Typically, a sudden onset, self-limited (resolves within 3 months), anterior, recurrent, unilateral uveitis. Most uveitis from any cause is anterior and 50% of those with a sudden onset of anterior uveitis are HLA B27 positive, most of whom have some form of spondyloarthropathy. Usually can be managed by topical corticosteroids.	HLA B27 typing; sacroiliac imaging to help diagnose associated spondyloarthritis
Idiopathic	The most common diagnosis in most series from a uveitis clinic. The term is used to mean a form of uveitis that does not fit a diagnostic niche. Other suggested terms include non-classifiable, undifferentiated, or primary (as opposed to secondary to another condition such as ankylosing spondylitis).	A diagnosis made when all other diagnoses have been excluded
Juvenile idiopathic arthritis associated uveitis	Characteristically a bilateral, insidious onset, chronic (lasting longer than 3 months), anterior uveitis, especially likely in	A +ANA is supportive but it is neither sensitive nor specific

	patients with JIA who have a pauci-articular, early onset, ANA positive form of arthritis	
Pars planitis	A relatively common form of uveitis that usually begins insidiously and causes inflammation in the vitreous humor resulting in prominent floaters. (However, the majority of patients with floaters do not have pars planitis.) The disease is named for the pars plana, an anatomic area immediately posterior to the ciliary body and a site where leukocytic concretions are found in this disease.	A clinical diagnosis based on the location of inflammation; occasionally associated with multiple sclerosis so a brain MRI is sometimes obtained.
Primary intraocular lymphoma	A very rare form of uveitis, usually occurring in individuals over 45, and presenting as bilateral cells in the vitreous humor, sometimes with subretinal infiltrates, and often in association with central nervous system lymphoma	Definitive diagnosis is made by characterization of malignant cells in the vitreous humor or on retinal biopsy.
Sarcoidosis	A form of uveitis with "promiscuous" or highly varied presentations ranging from anterior uveitis to retinal vasculitis with or without chorioretinal lesions. Ocular inflammation and pulmonary disease are the two most common initial manifestations of sarcoidosis and it accounts for a relatively common systemic illness among patients in a uveitis clinic.	Chest computed tomography is the most sensitive test. Biopsy is rarely required if symmetric hilar or mediastinal adenopathy is present. ACE, lysozyme, and interleukin-2 receptor have questionable specificity and limited sensitivity.
Syphilis	Late secondary or tertiary syphilis are in the differential diagnosis for any patient labelled as having "idiopathic" uveitis.	FTA or comparable test is preferred. RPR can be negative in up to 40% of patients with uveitis secondary to syphilis.
Tubulo-interstitial nephritis with uveitis	A form of uveitis that is typically sudden onset, bilateral and	Elevation of urine beta$_2$ microglobulin is a sensitive way

	mostly anterior with a variable amount of vitreous humor inflammation. The disease has an extremely strong association with HLA DRB1*0102. Patients with TINU are typically systemically ill with fever, myalgias and arthralgias as well as a markedly elevated sedimentation rate.	to support the diagnosis. Renal biopsy is definitive but often not required. Serum blood urea nitrogen or serum creatinine have limited sensitivity.
Tuberculosis	A rare form of uveitis in the United States, but a very common form of uveitis in countries such as India or Saudi Arabia. Also a very difficult diagnosis to confirm as the organism is rarely cultured from the eye. The diagnosis should be considered if the patient has a risk factor for TB (such as being born outside the US or a history of incarceration) or if the illness does not respond to immunosuppression such as oral corticosteroids.	Culture is the ideal confirmatory test, but it is often negative. PCR, when available, is a good alternative. Interferon gamma release assays and skin test responses are useful in confirming exposure to tuberculosis, but neither establishes if active infection is present.
Vogt-Koyanagi-Harada syndrome	An autoimmune form of uveitis with the triggering antigen putatively being tyrosinase. Patients are almost always either Asian, native American, or Spanish speaking. The uveitis is a bilateral, severe panuveitis with serous retinal elevation. Additional symptoms may include headache, meningismus, eight nerve abnormalities, and vitiligo.	Fluorescein angiography and/or optical coherence tomography should show characteristic serous elevation of the retina.
Whipple's disease	A rare but treatable cause of uveitis in association with arthritis. The presentation in the eye is generally cells in the vitreous humor.	Identification of the causative organism, *Tropheryma whippeli*, in the vitreous humor or elsewhere

Table 4. Forms of Uveitis of Potential Special Interest to Rheumatologists. Abbreviations: ACE=angiotensin converting enzyme; FTA=fluorescent treponemal antibody; RPR=rapid plasma reagin; PCR=polymerase chain reaction.