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# Retinal Manifestations in Juvenile Lupus Erythematosus

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## AIM

Juvenile systemic Lupus Erythematosus (JLE) is a multifaceted systemic autoimmune disease with frequent ocular manifestations. This study aims to review the reported JLE retinal manifestations comprehensively.

## METHODS

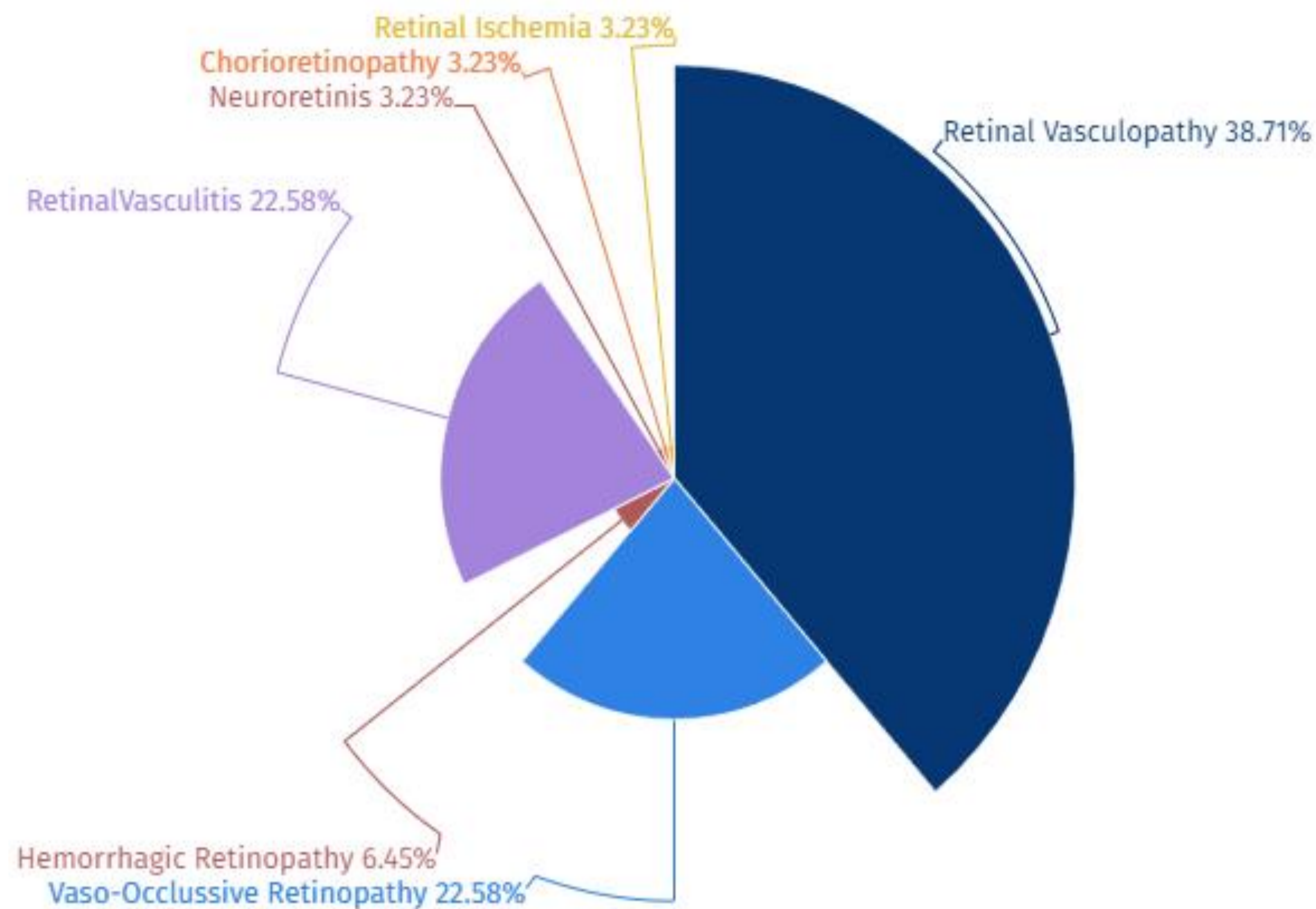
- A systematic search of the MEDLINE database for identification of relevant observational studies, case series and reports with retinal abnormalities in JLE patients from 1/2000 until 5/2023 was performed
- The collected data were analyzed to summarize retinal findings and their association with disease activity and prognosis

## RESULTS (1)

- 19 articles met the inclusion criteria, including 963 cases of JLE, among which 31 cases with retinal manifestations
- **Retinal involvement in JLE** manifested as:
  - Vaso-occlusive retinopathy
  - Retinal vasculopathy with cotton wool spots, Roth spots and hemorrhages
  - Retinal vasculitis
  - Chorioretinopathy
  - Neuroretinitis
  - Hemorrhagic retinopathy
  - Retinal ischemia
- These findings can cause significant visual impairment if left without treatment

frequency  
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Publication	Study Design	JLE cases	Retinal manifestation	Age (years ± SD)	Retinal manifestation
Dominguez et al (2022)	Case report	1	1	15	Retinal vasculitis
Wen et al (2021)	Case report	1	1	12	Hemorrhagic retinopathy due to thrombocytopenia
Guleria et al (2020)	Case report	2	2	9 and 8	Retinal vasculopathy (cotton-wool spots, occlusive micro-angiopathy)
Huang et al (2020)	Case report	1	1	11	CRAO/ CRVO with diffuse retinal hemorrhages and macular edema.
Páramo et al (2019)	Case report	1	1	14	Combined CRAO/ CRVO in the left eye and vitreous hemorrhage
Zhang et al (2017)	Case based review	1	1	15	Bilateral Hemorrhagic Retinopathy with Roth Spots
Korematsu et al (2014)	Case report	1	1	15	CRVO
Donnithorne et al (2013)	Case report	2	2	16 and 13	Retinal vasculitis and BRAO, non-arteritic ischemic optic neuropathy, retinal hemorrhage, cotton wool spots
Sguigna et al (2021)	Case report.	1	1	15	Asymptomatic retinal vasculopathy
Dorransoro et al (2023)	Case report	1	1	15	Bilateral neuroretinitis, bilateral papilla edema with partial macular star
Gawdat et al (2018)	Cross-sectional	40	7	13±2.8	Retinal vasculopathy
Deaner et al (2020)	Case report	1	1	15	Vaso-occlusive retinopathy, cotton wools spots
Gelman et al (2014)	Case report	1	1	12	Vaso-occlusive retinopathy
Almeida et al (2010)	Case report	2	2	15 and 20	Retinal vasculitis- associated with VZV infection
Alhassan et al (2021)	Case report	1	1	14	Retinal vasculitis and panuveitis
Firl et al (2020)	Case report	1	1	12	Retinal vasculitis associated with VZV, floaters, extensive peripheral vascular sheathing and retinal capillary nonperfusion
Fischer et al (2016)	Case report	1	1	14	Bilateral chorioretinopathy with transitory visual impairment
Kahwage et al (2017)	Retrospective cohort	852	1	-	Retinal ischemia/Uveitis
Al-Mayouf et al (2013)	Cross-sectional	52	4	11.3	Retinal vasculopathy



## RESULTS (2)

Retinal manifestations in JLE are caused by:

- **Antiphospholipid antibodies and immunoglobulins** accumulation in the vascular wall resulting in luminal narrowing
- **Active vasculitis** that may disrupt the vascular endothelium, exposing phospholipids as potential antigenic stimuli for antiphospholipid antibody production or as binding targets, triggering inflammatory pathways and thrombosis
- **Thrombocytopenia** and systemic hypertension
- **Treatment agents' toxicity and immunosuppression**

- The **presence and severity of retinal involvement often correlate** with systemic disease activity and may indicate disease progression
- Retinal involvement can occur **at any stage of the disease and may vary** in severity
- Three studies reported retinal involvement as the **initial presentation of JLE**

## CONCLUSIONS

- Our review underscores the imperative for clinical awareness about retinal manifestations in JLE
- Regular ophthalmic evaluation and close collaboration between rheumatologists and ophthalmologists are essential
- Further research is warranted to elucidate the pathogenesis of retinal involvement in JLE in order to develop targeted therapies to minimize ocular complications and improve long-term prognosis for those patients

## REFERENCES

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