

BACKGROUND

Familial exudative vitreoretinopathy (FEVR) is a hereditary disorder of peripheral retinal vascular development driven by impaired *Norrin* / *Frizzled-4* (*Wnt*) signalling. Mutations in **LRP5**, **FZD4**, **NDP**, **TSPAN12**, **KIF11** and **ZNF408** disrupt retinal angiogenesis, producing avascular periphery, neovascularisation, vitreoretinal traction, exudation and ultimately retinal detachment.

Pendergast & Trese stage 1–5 disease guides treatment: observation/laser early; **scleral buckle (SB)** for milder tractional / rhegmatogenous RD; **pars plana vitrectomy (PPV)** — often with lensectomy and silicone oil — for advanced detachments.

Surgical evidence is fragmented across small case series with variable reporting.

AIM

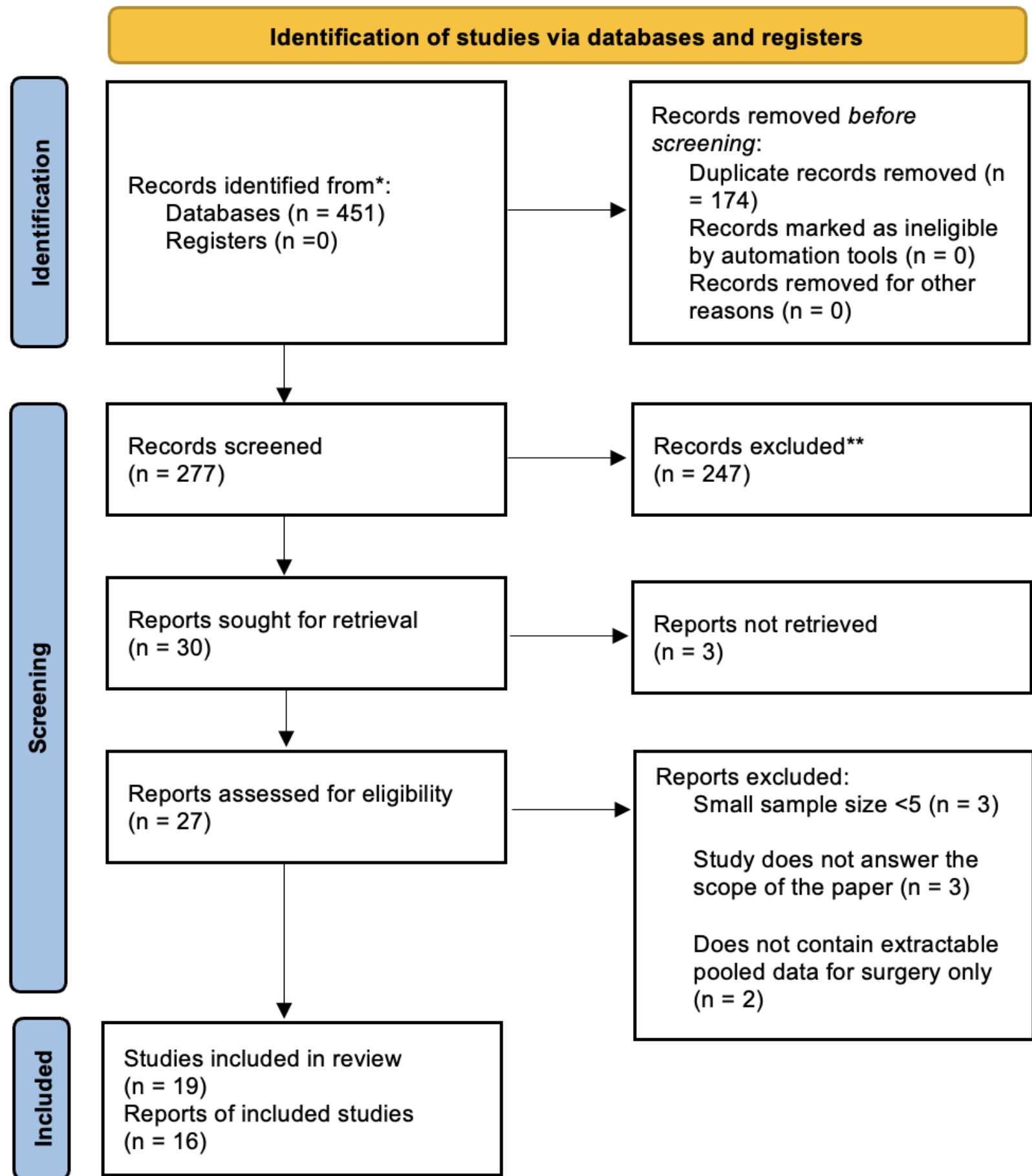
To systematically review and meta-analyse the anatomical and functional outcomes of vitreoretinal surgery for FEVR-associated retinal detachment.

METHODS

Conducted per **PRISMA 2020** (PROSPERO CRD420251170193). PubMed, EMBASE, Ovid, Medline and Cochrane CENTRAL searched from inception to Feb 2026.

- **Population:** patients with FEVR-associated retinal detachment (≥5 eyes)
- **Intervention:** scleral buckle, pars plana vitrectomy, or combined surgery
- **Outcomes:** retinal reattachment rate, postoperative BCVA, complications, reoperation
- **Risk of bias:** JBI Critical Appraisal Checklist for Case Series
- **Synthesis:** single-arm proportion meta-analysis (REML, HKSJ adjustment) — meta package, R

PRISMA 2020 SELECTION



STUDY OVERVIEW

19
studies included

682
eyes operated on

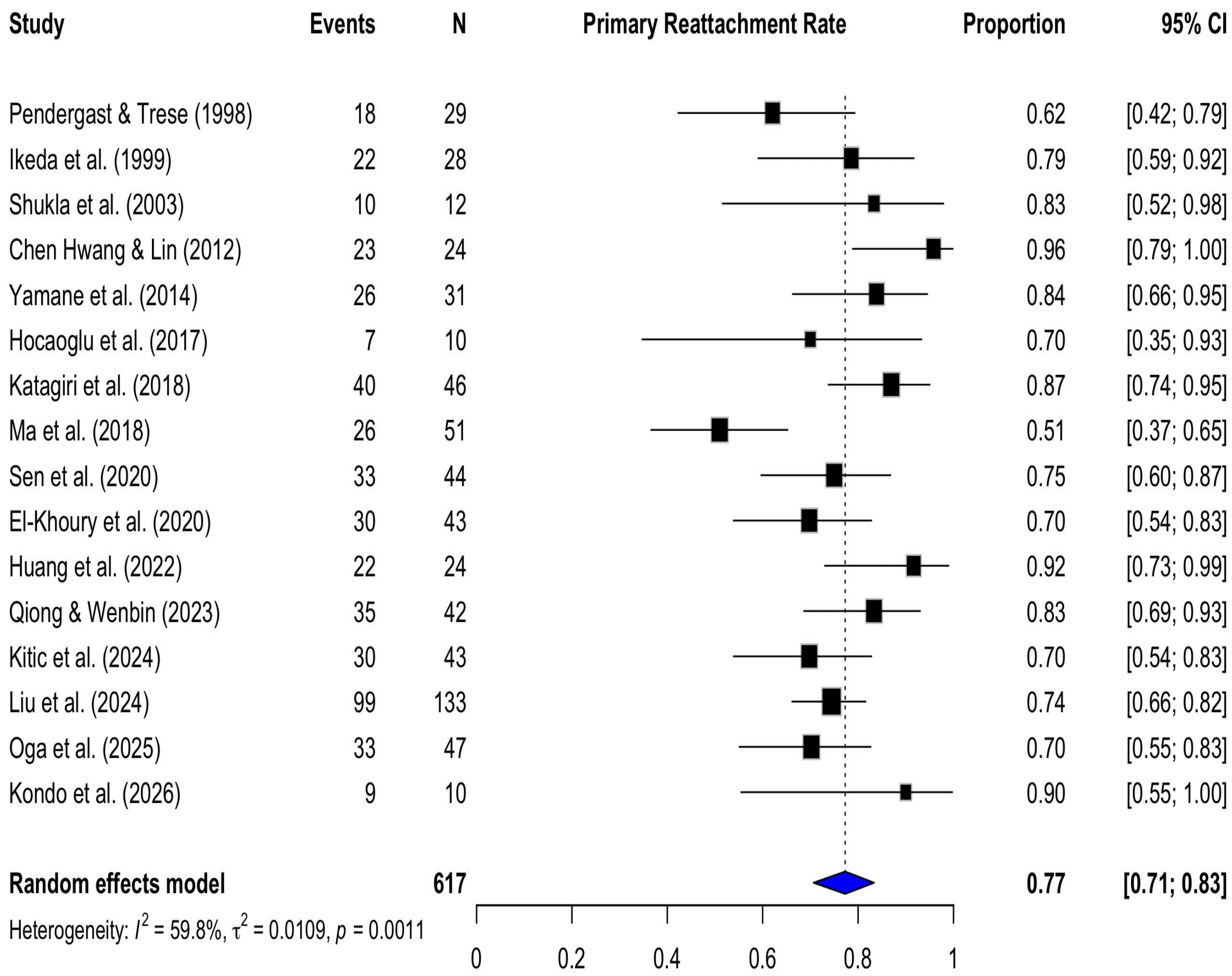
0.77
pooled reattachment

0.24
pooled reoperation

All 19 studies were retrospective or observational case series. Surgical interventions: PPV alone, SB alone, or combined SB + PPV (often with lensectomy). Predominantly paediatric cohorts; FEVR stages 1–5 represented.

RESULTS — PRIMARY REATTACHMENT

Pooled across 16 studies: **0.77** (95% CI 0.71–0.83; $I^2 = 59.8\%$; $p = 0.0011$).

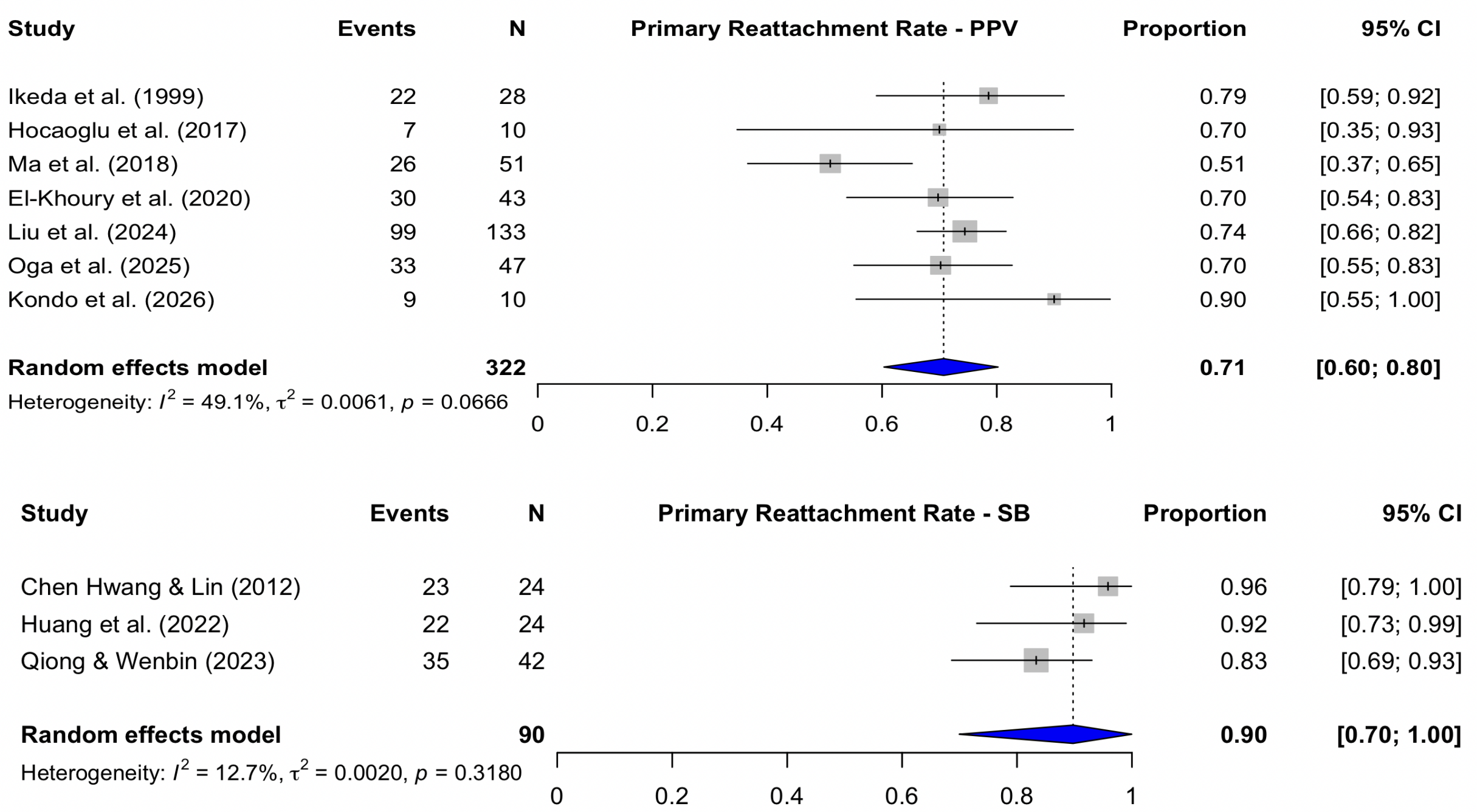


Reoperation rate **0.24** (5 studies; 95% CI 0.08–0.45; $I^2 = 84.7\%$) reflects the complexity of these eyes.

SUBGROUP — SB vs PPV

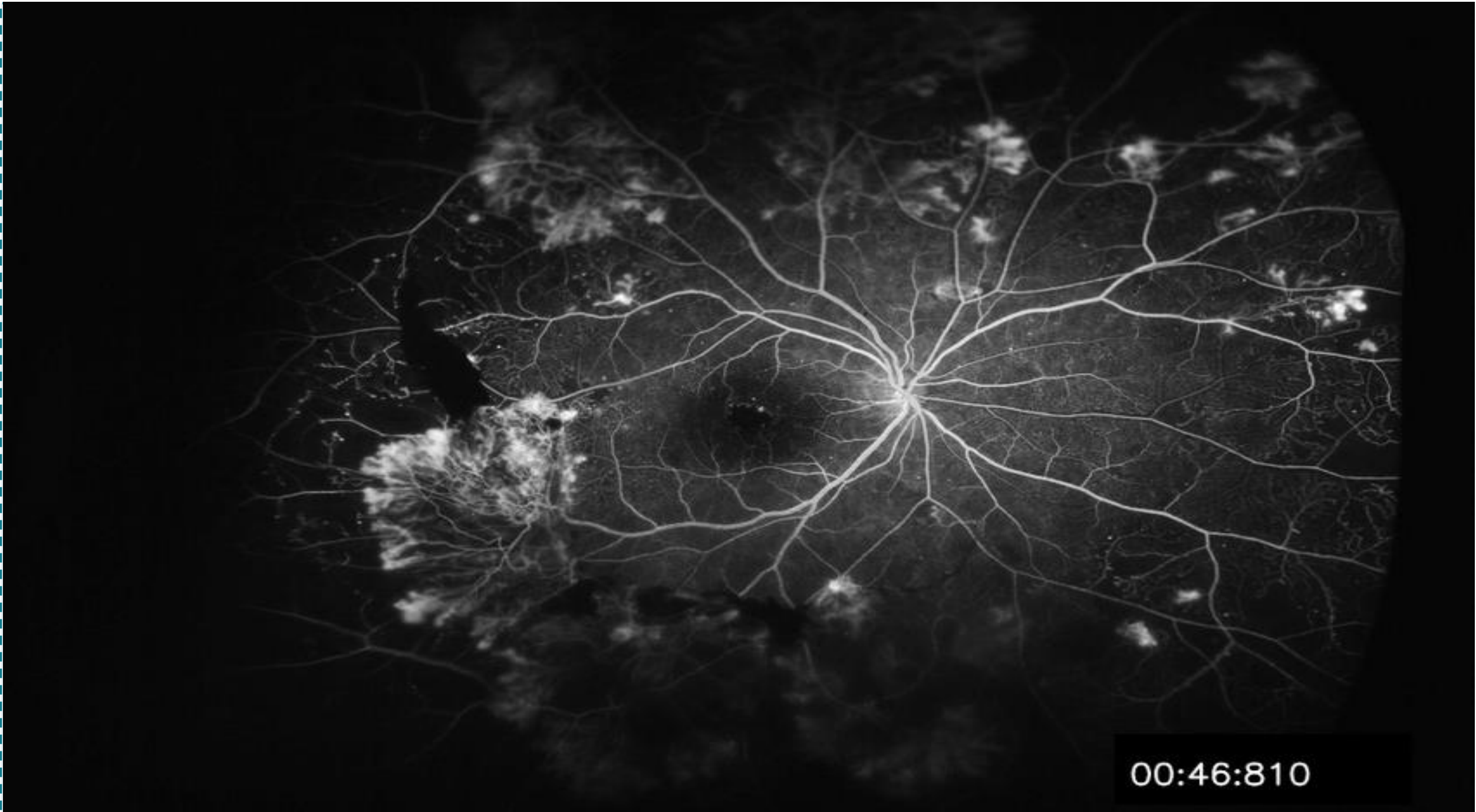
Scleral buckle (3 studies): **0.90** (95% CI 0.70–1.00; $I^2 = 12.7\%$).

Pars plana vitrectomy (8 studies): **0.71** (95% CI 0.60–0.80; $I^2 = 49.1\%$).



SB shows higher numerical reattachment, but overlapping CIs and small SB sample (n = 3) call for cautious interpretation. PPV is preferentially used in advanced disease.

VISUAL & STAGE-DEPENDENT OUTCOMES



- **Stage-dependent success** (Oga 2025): Stage 3 100% · Stage 4 77.4% · Stage 5 12.5%.
- **FA grade** was the only independent predictor of reattachment (OR 9.0, 95% CI 1.2–70; $p = 0.04$).
- **BCVA improved** in most reporting studies — Agrawal 2022: 1.42 → 0.60 logMAR ($p < 0.001$); Huang 2022: 1.08 → 0.45 (79% improved).
- Combined SB + PPV may achieve 91–100% reattachment in fibrovascular proliferation cases.

DISCUSSION

- Surgery is generally effective: pooled reattachment **0.77** — but moderate heterogeneity ($I^2 = 59.8\%$) reflects diverse stages, techniques and reporting.
- **Stage-tailored escalation:** SB for milder Stage 2–3 disease; PPV for Stage 4–5 with complex traction; lensectomy + silicone oil for Stage 5.
- **FA grade** may serve as a prognostic biomarker — vascular activity, not anatomical stage alone, should inform surgical planning.
- Combined SB + PPV is increasingly favoured for fibrovascular proliferation across ≥2 quadrants.
- Genetic counselling and FA screening of relatives identifies subclinical carriers eligible for prophylactic laser.

LIMITATIONS

- Only retrospective case series / cohorts — no randomised comparisons.
- Heterogeneous BCVA reporting precluded quantitative pooling of visual outcomes.
- Detachment mechanisms (tractional vs rhegmatogenous vs exudative) not stratified across most studies.
- Egger's test $p = 0.271$ — no clear publication bias, but limited power ($n = 19$).

CONCLUSION

Vitreoretinal surgery for FEVR-associated RD achieves **favourable primary reattachment rates**, with outcomes strongly influenced by disease stage. SB suits early/uncomplicated detachments; PPV (± SB ± lensectomy ± silicone oil) is required for advanced disease. **Standardised reporting and prospective comparative studies are needed.**

KEY REFERENCES

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