

# Primary Cardiac Angiosarcoma: A Systematic Review of Clinical Presentation, Management, and Outcomes

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## INTRODUCTION & AIM

Primary cardiac angiosarcoma (PCA) is an exceptionally rare and aggressive malignancy with a poor prognosis. This systematic review synthesizes the current evidence on the epidemiology, clinical presentation, diagnostic approaches, management strategies, and outcomes of PCA to guide clinical practice.

## METHOD

We conducted a systematic review of studies published from inception to January 2026. Our search was performed exclusively in the PubMed electronic database for English-language studies. The review encompassed various study designs, including case reports, case series, cohort studies, and systematic reviews, to comprehensively capture the available evidence on this rare condition. A total of 49 studies were selected for final inclusion from an initial 710 identified records after a rigorous, multi-stage screening process.

## RESULTS & DISCUSSION

49 studies were included. PCA predominantly affected middle-aged adults (median age 40s–50s) with a significant male predominance ( $\approx 69\%$ ). The vast majority of tumors (92%) originated in the right atrium. Clinical presentation was often non-specific, including pericardial effusion, heart failure symptoms, and hemoptysis. Diagnosis relied on multimodality imaging (Echocardiography, Cardiac MRI, CT) and was confirmed by biopsy and immunohistochemistry (positive for CD31, CD34). Management was multimodal, with complete surgical resection (R0) being the cornerstone of potentially curative treatment. Adjuvant or neoadjuvant chemotherapy (often paclitaxel-based) and radiotherapy were frequently employed. Despite treatment, overall prognosis remained poor, with a median survival of approximately 6–26 months.

Fig. 1. Demographic Characteristics of Included Patients (N = 49 Studies)

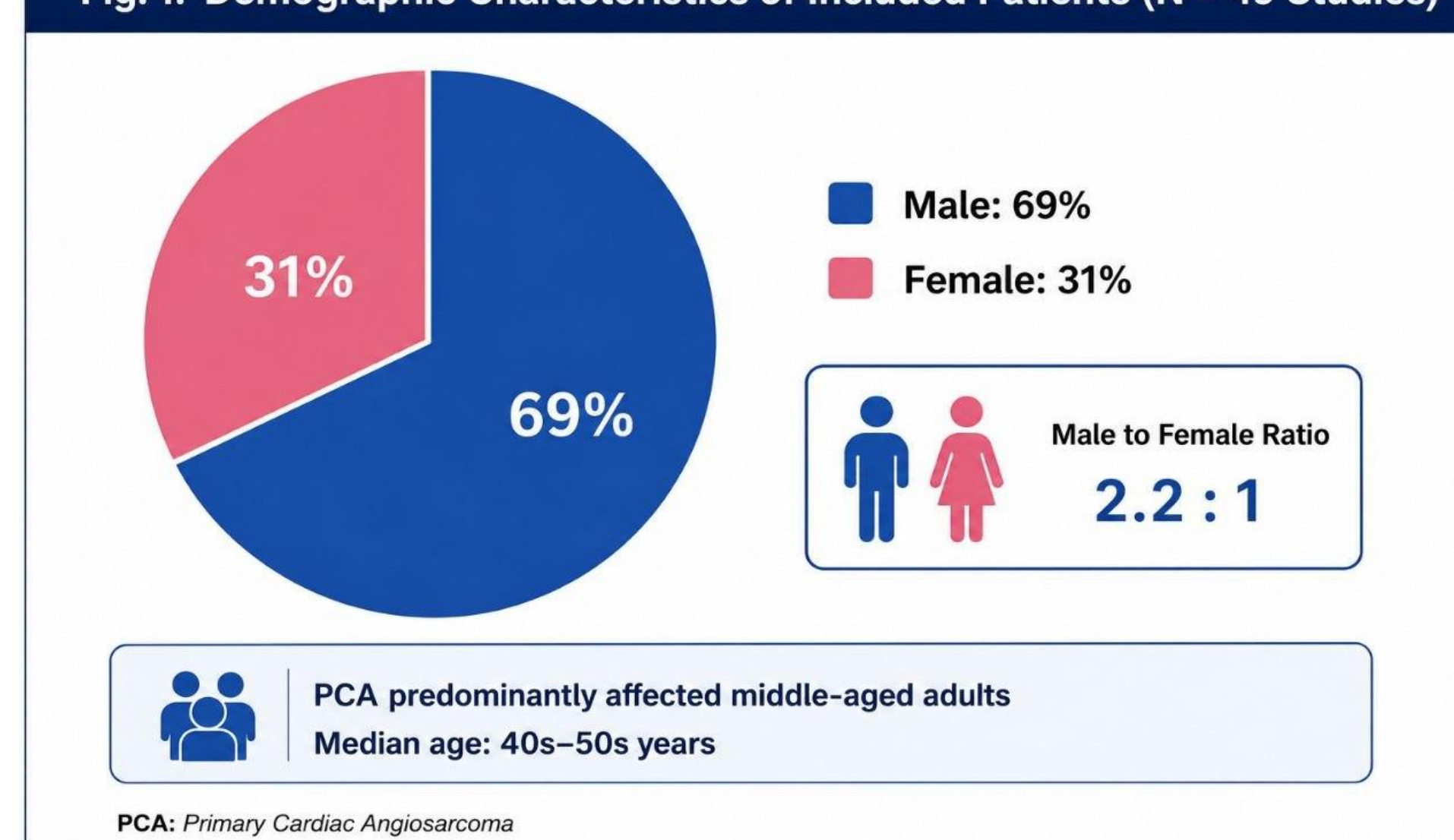


Fig. 2. Primary Tumor Location

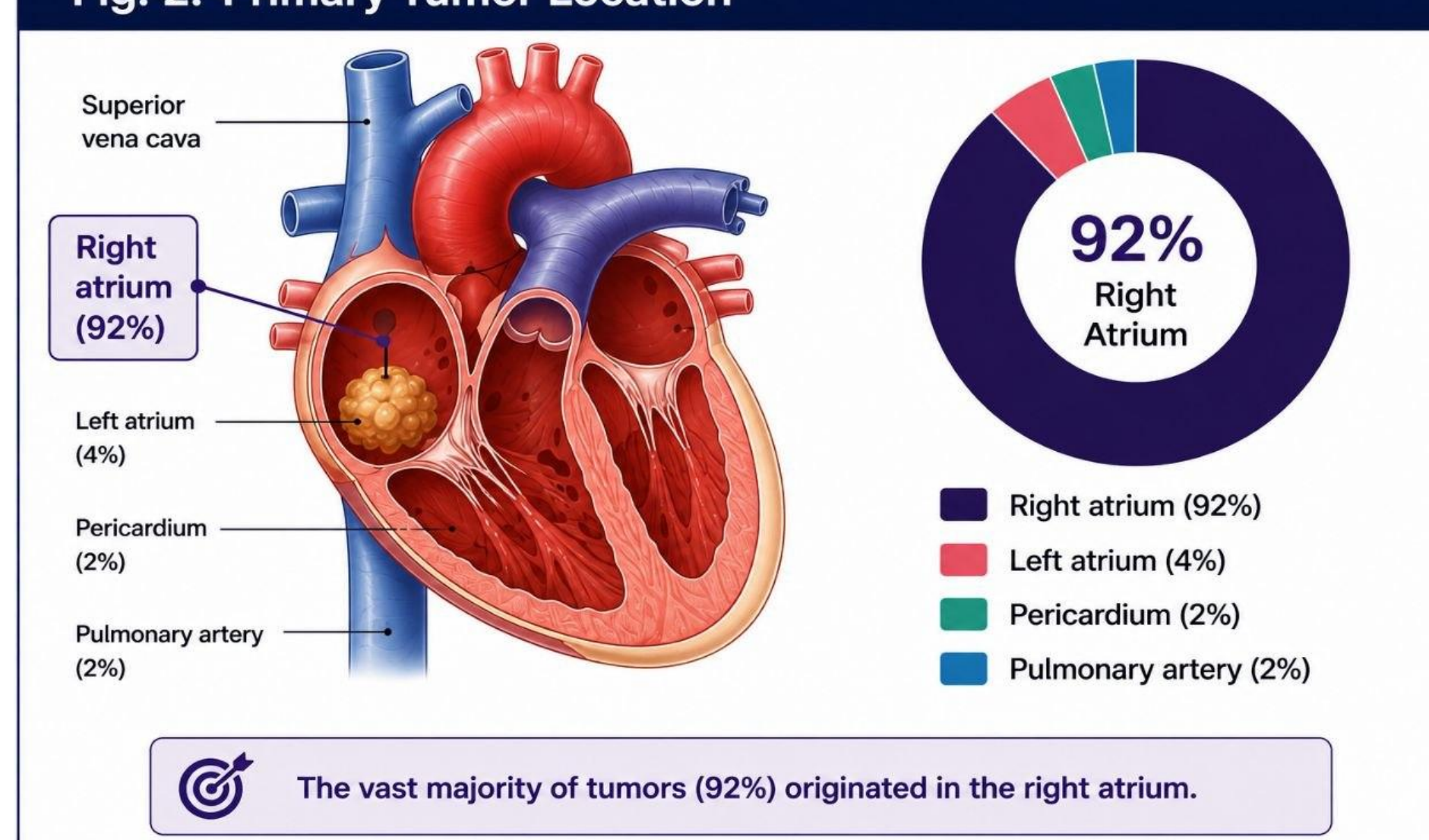


Fig. 3. Clinical Presentation at Diagnosis (N = 49 Studies)

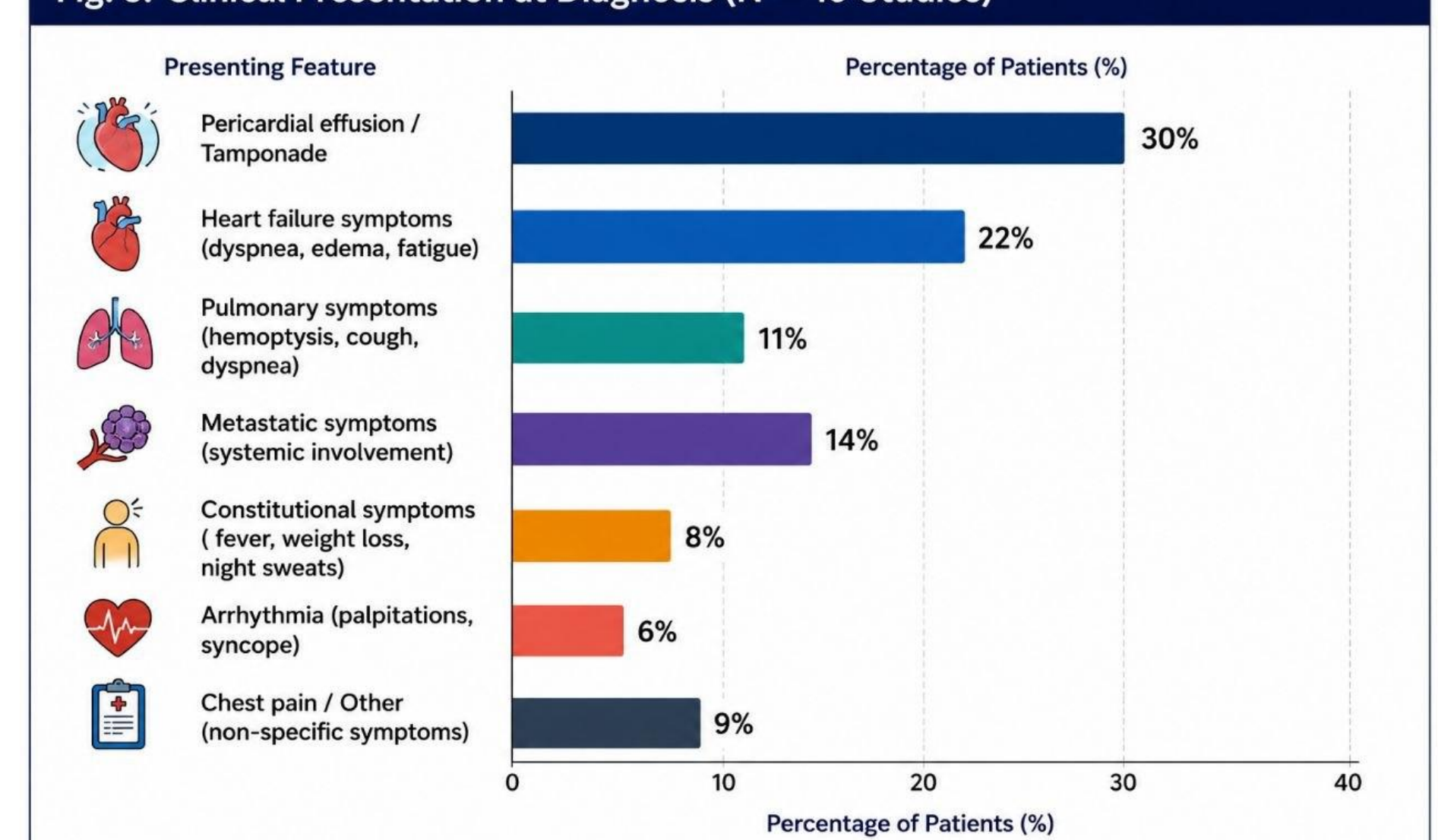


Fig. 4. Diagnostic Approach and Key Findings

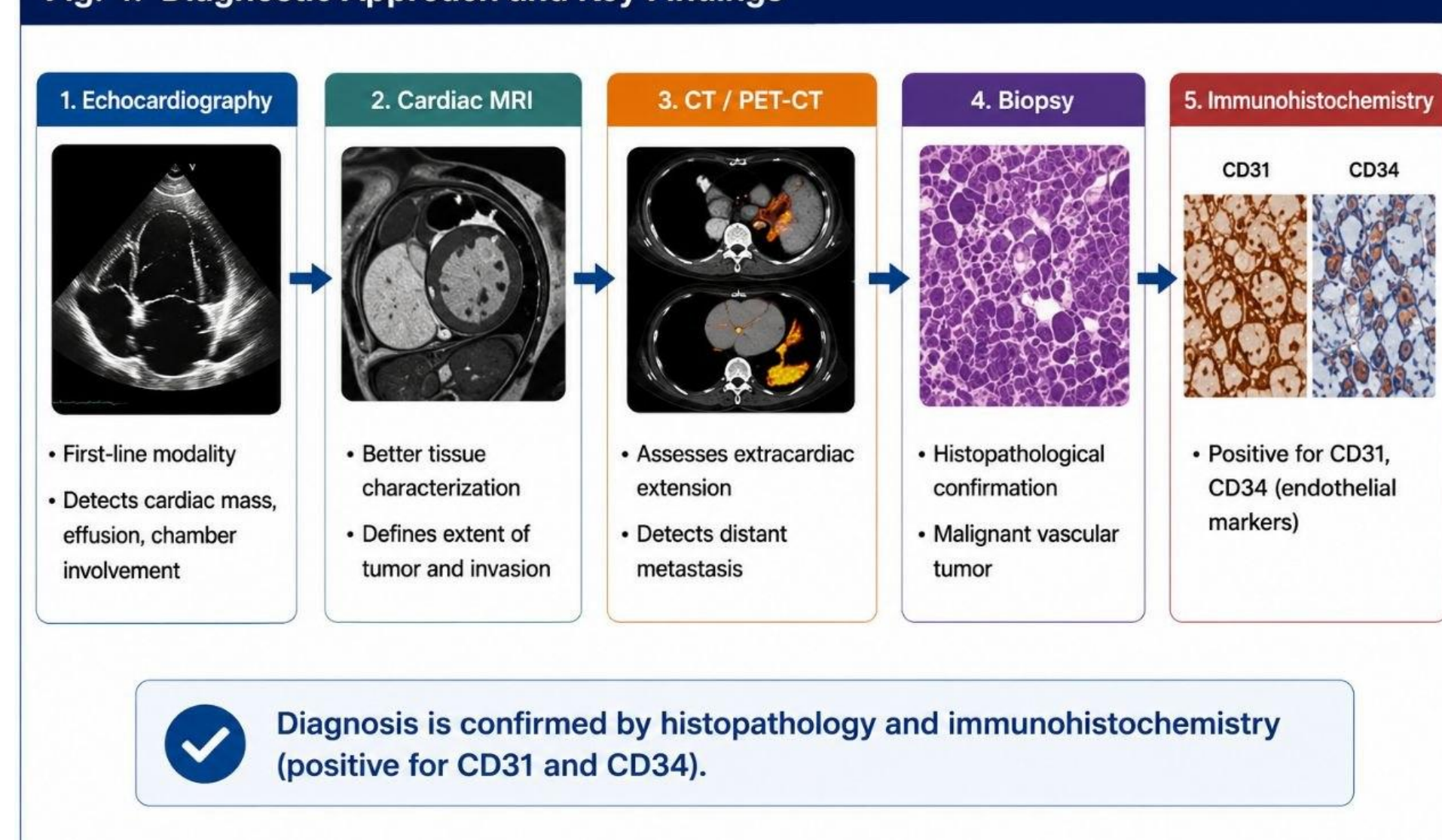


Fig. 5. Treatment Modalities and Outcomes (N = 49 Studies)

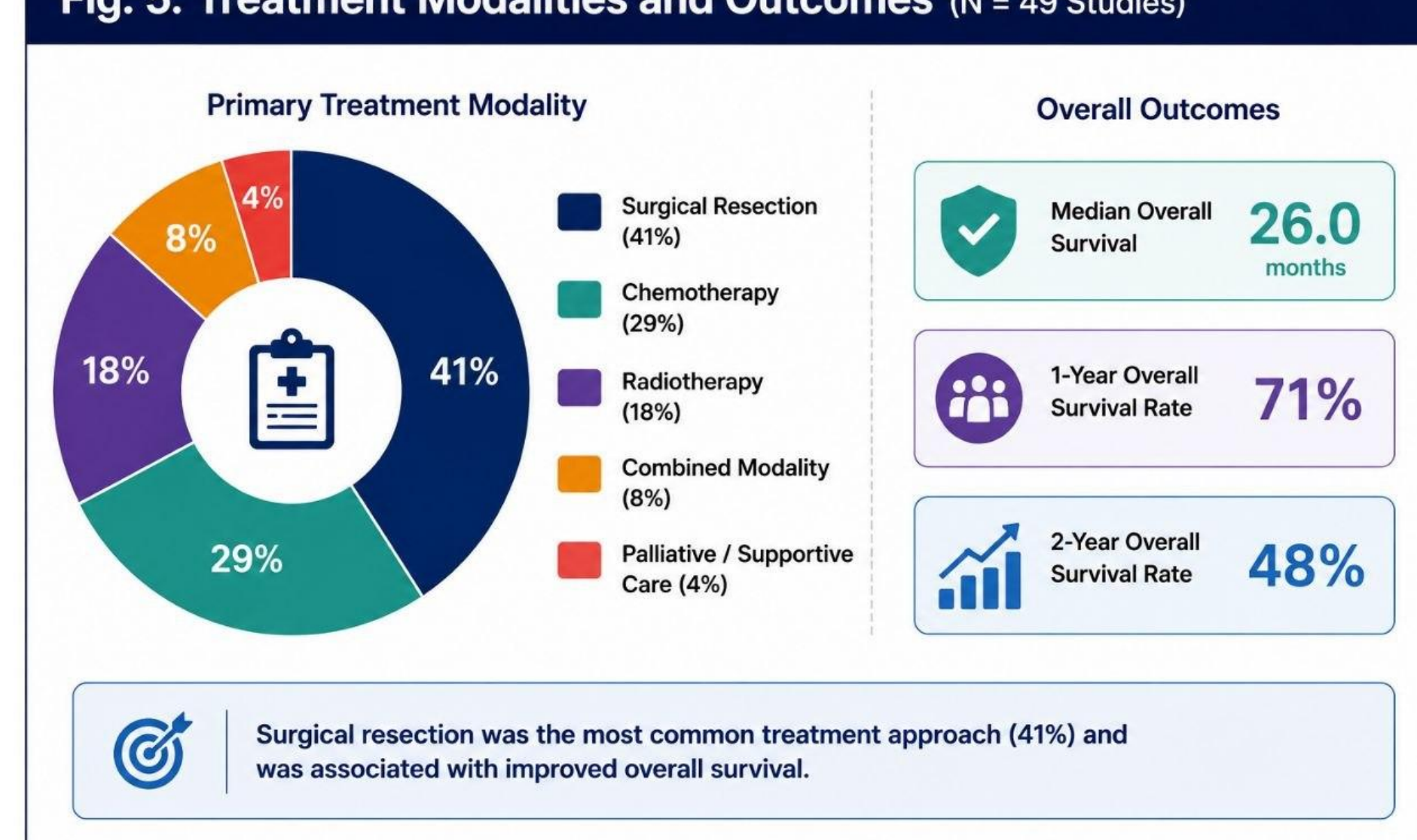
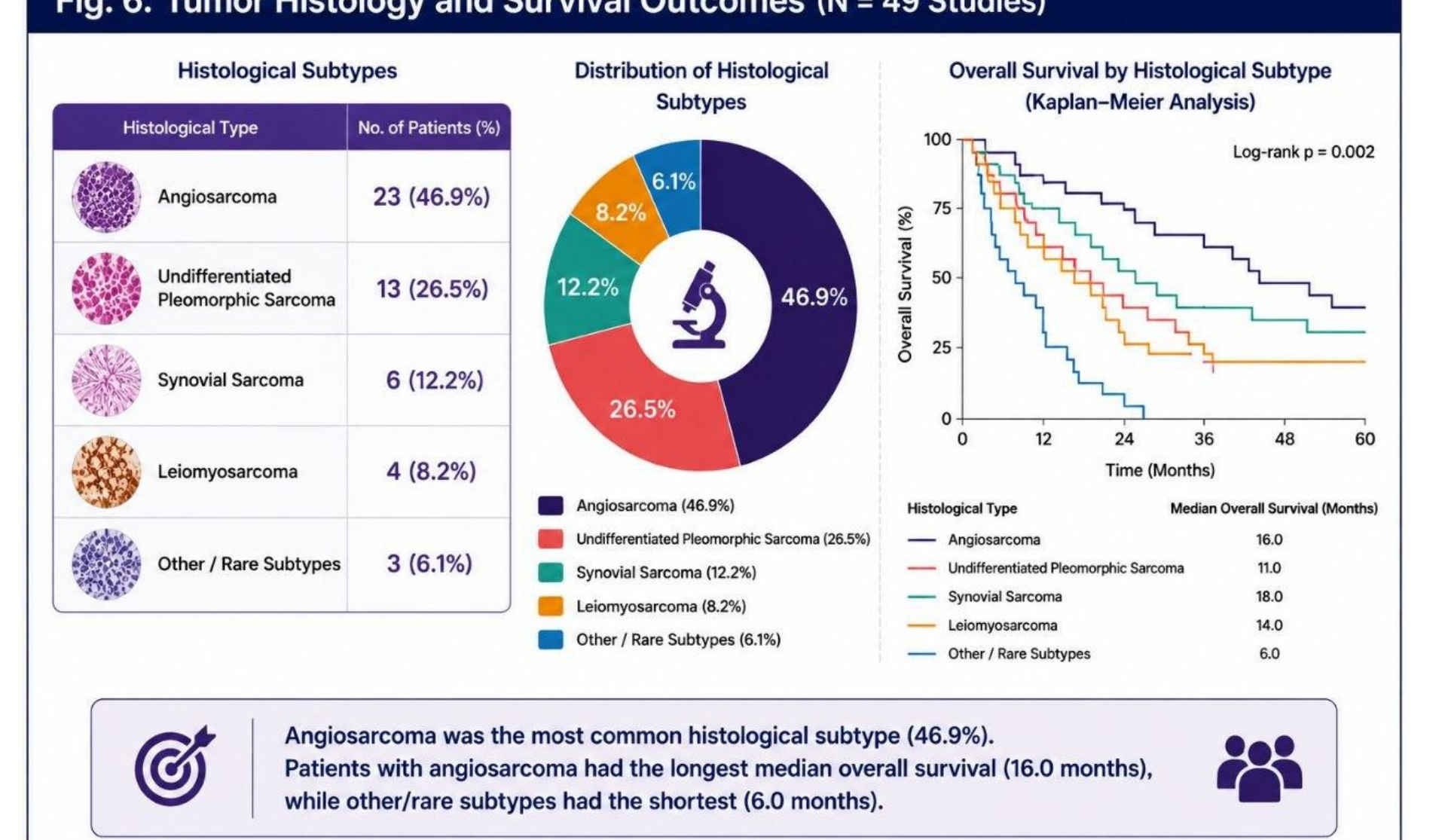


Fig. 6. Tumor Histology and Survival Outcomes (N = 49 Studies)



## CONCLUSIONS

PCA is a devastating disease often diagnosed at an advanced stage. A high index of suspicion in patients with right-sided cardiac masses or unexplained pericardial effusion is crucial. Early diagnosis via multimodality imaging, prompt biopsy, and an aggressive, multimodal treatment strategy offer the best chance for improved survival. International collaboration is urgently needed to establish standardized guidelines.