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Pulmonary Artery Aneurysm in Behçet's Disease and Hughes–Stovin Syndrome: A Systematic Review of Prevalence, Mortality, and Geographic Variation

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Abstract

Background: Behçet's disease (BD) is a multisystemic vasculitis characterized by inflammation of vessels of all calibers, with vascular involvement representing one of its most severe manifestations. Among arterial complications, pulmonary artery aneurysm (PAA) is the most distinctive and life-threatening lesion. Despite its rarity, PAA contributes disproportionately to morbidity and mortality in BD.

Objectives: This systematic review aimed to determine the prevalence, clinical spectrum, and outcomes of PAA in BD, and to contextualize Hughes–Stovin syndrome (HSS) as a related entity within the same vasculitic continuum.

Methods: Following PRISMA 2020 guidelines, a comprehensive search was conducted in PubMed/MEDLINE up to October 2025 using terms related to Behçet's disease, pulmonary artery aneurysm, and Hughes–Stovin syndrome. Denominator-defined cohorts, descriptive pulmonary BD series, and HSS compilations were included. Data on study design, sample size, imaging method, prevalence, and mortality were extracted and summarized descriptively.

Results: Sixty-two publications were identified, of which fourteen met the inclusion criteria. Two denominator-based studies provided prevalence estimates between 0.16% and 1.1%, while several radiologic and clinical series confirmed PAA as the predominant pulmonary vascular manifestation of BD. Mortality historically approached 50% in early series but has declined to approximately 25–30% with modern immunosuppressive and biologic therapies. PAA occurs mainly in young men and is most frequently reported in Turkey and the Middle East, paralleling the geographic distribution of BD. HSS remains exceedingly rare, with fewer than 60 cases documented worldwide, and is now regarded as an incomplete or monosymptomatic form of BD.

Conclusions: Pulmonary artery aneurysm is an uncommon but pathognomonic feature of Behçet's disease, affecting roughly 1% of patients and associated with high but improving mortality. Advances in imaging and immunosuppressive therapy have enhanced detection and survival. The close overlap between HSS and BD supports their inclusion within a shared vasculitic spectrum. Multicenter registries and standardized imaging criteria are needed to clarify regional disparities and long-term outcomes.

Keywords: Behçet's disease, pulmonary artery aneurysm, vascular involvement, Hughes–Stovin syndrome, vasculitis, prevalence, mortality, computed tomography angiography, biologic therapy, epidemiology

Introduction

Behçet's disease (BD) is a chronic, relapsing, multisystem vasculitis that affects arteries and veins of all calibers, being categorized among the variable-vessel vasculitides [1]. It presents with mucocutaneous, ocular, neurologic, gastrointestinal, and vascular manifestations whose prevalence varies geographically [2]. The disease is most common along the historic Silk Road, including Turkey, Iran, and East Asia, where prevalence rates range from 20 to 420 per 100,000 inhabitants [3, 4]. Vascular

involvement,sometimes referred to as angio-Behçet, occurs in up to 40% of patients [5] and represents one of the main causes of morbidity and mortality [6]. Among arterial lesions, pulmonary artery aneurysm (PAA) stands out as a pathognomonic and life-threatening complication [7, 8].

The earliest large clinical series describing pulmonary vascular involvement in BD were reported in Turkey, where vascular forms of the disease are particularly frequent [9]. In the landmark cohort by Hamuryudan *et al*. (1994) [10], 24 PAA cases were identified among 2,179 patients with BD, corresponding to a prevalence of 1.1%. Radiologic evaluation by Numan *et al*. (1994) [11] demonstrated that aneurysms were often multiple, bilateral, and associated with arterial occlusions. Before the advent of spiral computed tomography (CT), most diagnoses relied on conventional angiography, limiting detection to central lesions [12]. With the introduction of CT pulmonary angiography (CTPA), the sensitivity for small or peripheral aneurysms increased, leading to broader recognition of pulmonary vascular disease in BD [13].

Several reviews have reinforced the dominance of PAA among thoracic vascular manifestations of BD [14, 15]. Erkan *et al*. (2001) [16] highlighted that hemoptysis and aneurysmal rupture were key determinants of mortality. Uzun *et al*. (2005) [17], in a cumulative analysis of pulmonary vasculitis in BD, confirmed that PAA is the most frequent pulmonary vascular lesion. Later, Seyahi *et al*. (2012) [18] studied 47 patients with pulmonary artery involvement (PAI) and found that 72% had aneurysms, whereas the remainder presented with isolated thrombosis.

Despite improved imaging, the true frequency of PAA among all BD patients remains uncertain due to referral and geographic biases [19]. Most denominator-defined cohorts originate in Turkey, Iran, or China—countries with the highest BD prevalence [3, 4, 20]. Western reports are rare and mostly anecdotal [21], raising questions about underdiagnosis or genuine geographic variation. Mortality historically approached 50% in older series [10], although outcomes appear to have improved with aggressive immunosuppression and modern imaging surveillance [18, 22].

A related condition, Hughes–Stovin syndrome (HSS), described by Hughes and Stovin (1959) [23], is characterized by recurrent thrombophlebitis and pulmonary artery aneurysms. Pathologically, HSS shares features with BD, leading many authors to consider it an incomplete or monosymptomatic form of BD [24, 25]. However, due to its rarity—fewer than 60 cases reported worldwide—no reliable frequency estimates exist [26].

Given these uncertainties, a PRISMA-based systematic review is warranted to synthesize all available literature providing denominator-based estimates of PAA prevalence and incidence in BD, and to contextualize HSS within the same vasculitic spectrum. This review aims to clarify the global epidemiological burden of pulmonary artery aneurysms in BD, identify regional heterogeneity, and highlight gaps for future population-level registry studies.

Methods

This systematic review was conducted in accordance with the PRISMA 2020 statement [27]. A comprehensive search was performed in PubMed/MEDLINE from database inception through 7 October 2025, using combinations of Medical Subject Headings (MeSH) and free-text terms related to *Behçet's disease* and *pulmonary artery aneurysms*. The primary search strategy included the terms *"Behçet"** or *"Behcet"** combined with *"pulmonary artery aneurysm," "pulmonary arterial aneurysm," "pulmonary vasculitis,"* or *"pulmonary involvement."* In addition, the term *"Hughes–Stovin"* was searched separately to identify studies and case reports concerning this related clinical entity [28]. Reference lists from relevant publications, including major narrative reviews and epidemiological studies [17, 29, 30], were screened manually to identify additional eligible sources. No language or publication-date restrictions were applied, and abstracts indexed in PubMed were considered if they provided original data [31].

Eligible studies were required to report original data on patients fulfilling internationally accepted diagnostic criteria for Behçet's disease, such as the 1990 International Study Group (ISG) or 2014 International Criteria for Behçet's Disease (ICBD) [32, 33].

Only studies that included a clearly defined denominator (total number of BD patients evaluated within a cohort or institutional database) and a corresponding numerator (number of BD patients with radiologically or angiographically confirmed pulmonary artery aneurysm) were included. Information on the study period, imaging methods, and patient follow-up was also required.

Case reports and small series that did not provide a BD denominator were excluded from the quantitative synthesis but were used qualitatively to contextualize natural history, morphology, and mortality. For Hughes–Stovin syndrome (HSS), given its extreme rarity and absence of denominator data, only cumulative case-based reviews and systematic compilations were analyzed [23, 26, 34].

All titles and abstracts were screened independently by two reviewers. Full texts were retrieved for potentially eligible studies, and discrepancies were resolved by consensus. A PRISMA flow diagram was prepared to illustrate the selection process [27].

Data extraction was performed using a standardized template including author, publication year, country, study setting, sample size, imaging modality, number of PAA cases, calculated frequency, and reported outcomes [10, 17, 18, 35]. Each eligible article was categorized as either a denominator-defined cohort (suitable for prevalence estimation), a pulmonary Behçet cohort (restricted to thoracic involvement and thus excluded from prevalence pooling), or an HSS series (summarized qualitatively).

For each study, the extracted variables comprised country and setting (to explore geographic heterogeneity), study period (to evaluate temporal trends), imaging era (angiography, early CT, or multidetector CT/CTPA), total number of BD patients, number of PAA cases, demographic characteristics, presenting symptoms—particularly hemoptysis—and mortality [6, 10, 12, 17, 18, 36]. Mortality was recorded as the proportion of deaths among patients with PAA when available. Data were entered into a master evidence table for descriptive synthesis.

The methodological quality of each included study was assessed qualitatively according to the principles of the Newcastle–Ottawa Scale (NOS) for nonrandomized studies [37]. Three major domains were considered:

Selection Bias, Related to the Representativeness of the BD Population and Diagnostic Criteria;
Ascertainment Bias, Reflecting Differences in Imaging Sensitivity Between Eras;
Outcome Bias, Referring to Selective Reporting of Deaths or Incomplete Follow-Up

Given the marked heterogeneity and small sample sizes, no numeric scoring system was applied. Referral bias was anticipated as most cohorts originated from tertiary rheumatology centers in Turkey and the Middle East [9, 10, 17, 19].

Because of heterogeneity in study design, sample size, and imaging methodology, a quantitative meta-analysis was not feasible. A descriptive synthesis was therefore performed, emphasizing frequency estimates, ranges, and regional patterns. Denominator-defined cohorts were prioritized for quantitative description, while pulmonary BD series and HSS reports were analyzed narratively to highlight clinical spectrum and prognosis [12, 17, 18]. Results were stratified by geographical region (Middle East/Turkey, Asia, Europe, and others) and imaging era to account for evolving diagnostic technology. Data consistency was verified independently by both reviewers before final synthesis.

Results

A total of 62 publications were retrieved through the PubMed search, of which 14 met the inclusion criteria after screening and full-text assessment. The remaining studies were excluded for lacking a defined denominator of Behçet's disease (BD) patients or for reporting only individual cases of pulmonary artery aneurysm (PAA). Of the 14 eligible articles, two provided denominator-based estimates suitable for prevalence calculation, eight were descriptive pulmonary BD cohorts, and four described cumulative reviews or compilations including cases of Hughes–Stovin syndrome (HSS). Table 1 sumamrizes cases of BD in which PAA was evaluated and Table 2 summarizes the included studies of HSS and its relationship with BD.

Table 1: Summary of studies reporting pulmonary artery aneurysm (PAA) in Behçet's disease.

Author, Year	Country / Region	Study Type / Period	Total BD patients (n)	PAA cases (n)	Prevalence (%)	Imaging method	Mortality (%)	Main findings / Notes	Ref.
Hamuryudan *et al*., 1994	Turkey	Retrospective cohort (1977–1993)	2,179	24	1.1	Angiography	~50	Multiple bilateral aneurysms; hemoptysis major cause of death	[10]
Numan *et al*., 1994	Turkey	Angiographic series	15	15	—	Angiography	53	31 aneurysms; 8 with associated thrombosis	[11]
Kural *et al*., 1999	Turkey	Hospital series	28	—	—	Angiography	43	Central lesions; high mortality	[12]
Tunacı *et al*., 1999	Turkey	Radiologic series	12	12	—	Helical CT	33	Partial regression under corticosteroids + cyclophosphamide	[40]
Uzun *et al*., 2005	Turkey	Cumulative review	598	—	—	Various	—	PAA as main pulmonary vascular lesion; male predominance	[17]
Çelik *et al*., 2015	Turkey	Multicenter review	—	207	0.16–1.10	CT / CT angiography	29	Improved detection with modern CT; variability across centers	[7]
Seyahi *et al*., 2012	Turkey	Prospective cohort	47	34	—	CT angiography	26	72% aneurysms, 28% isolated thrombosis	[18]
Yılmaz *et al*., 2021	Turkey	Multicenter series	61	11	18	CT angiography	23	Intracardiac thrombi in 20% of patients	[41]
Zhang *et al*., 2015	China	Hospital series	15	9	60	Helical CT	20	Hemoptysis as predominant symptom	[42]
Naguib *et al*., 2015	Egypt	Clinical series	15	5	33	CT angiography	—	Severe cases; response to immunosuppression	[43]
Davatchi *et al*., 2015	Iran	National registry	—	—	2–3	—	—	Vascular involvement in 25% of BD; PAA in 2–3%	[44]
Yazgan *et al*., 2021	Turkey	Radiologic review	—	—	—	CT / CT angiography	—	Peripheral and central lesions; non-aneurysmal findings common	[45]

Angio-TC = Computed tomography angiography; BD = Behçet's disease; CT = Computed tomography; CTPA = Computed tomography pulmonary angiography; HSS = Hughes–Stovin syndrome; PAA = Pulmonary artery aneurysm.

Table 2: Summary of studies reporting Hughes–Stovin syndrome (HSS) and its relationship with Behçet's disease.

Author, Year	Country / Region	Study Type / Period	Total HSS Cases (n)	Male (%)	Mean Age (years)	Pulmonary Lesions	Associated Thrombosis	Mortality (%)	Main Findings / Interpretation	Ref.
Khalid & Saleem, 2011	USA	Systematic review	40	95	32	Multiple pulmonary artery aneurysms	100	42	Classic sequence of deep vein thrombosis followed by aneurysm formation; clinical overlap with vascular BD	[23]
Ciancio *et al*., 2014	Italy	Case report and literature review	—	—	—	Present	Present	—	Histopathologic similarities to BD; supports shared vasculitic mechanism	[24]
Mohammad *et al*., 2022	Sweden	Systematic review	55	87	31	Variable, mostly bilateral aneurysms	Frequent	33	Proposes HSS as incomplete or limited form of BD; fewer mucocutaneous features	[60]
Elmaghraby *et al*., 2023	Egypt	Systematic review	60	88	30	Bilateral and multiple aneurysms	Frequent	35	Updated global synthesis (2023); reinforces overlap with BD	[61]
Gatto *et al*., 2023	Italy	Narrative review	—	—	—	Variable	Variable	—	Highlights shared vasculitic spectrum linking BD and HSS	[62]

BD = Behçet's disease; HSS = Hughes–Stovin syndrome; PAA = Pulmonary artery aneurysm; USA = United States of America; — = Data not available.

The earliest denominator-defined study remains the report by Hamuryudan *et al.* (1994) [10], who evaluated 2,179 consecutive BD patients and identified 24 with PAA, corresponding to a prevalence of approximately 1.1%. This cohort, originating from a tertiary referral center in Istanbul, Turkey, included predominantly young men with severe vascular disease. Mortality was nearly 50%, with most deaths resulting from hemoptysis secondary to aneurysmal rupture. The aneurysms were generally multiple, bilateral, and located in the lower lobes.

A second denominator-based estimate can be inferred from Turkish multicenter syntheses summarized by Çelik *et al.* (2015) [7], which reported a prevalence range of 0.16% to 1.10% among national BD registries, depending on diagnostic criteria and imaging era. The authors emphasized that the variability was influenced by improved detection through CT angiography and the increased recognition of small peripheral aneurysms after 2000. Radiologic series have provided complementary insight into lesion morphology and progression. In the angiographic study by Numan *et al.* (1994) [11], 15 BD patients with pulmonary involvement exhibited 31 aneurysms, of which 18 were multiple and 8 were associated with thrombosis. Subsequent radiologic follow-up demonstrated partial or complete regression of some lesions under corticosteroid and cyclophosphamide therapy, suggesting that immunosuppression could alter the course of vascular inflammation [39].

Further imaging-based analyses confirmed these findings. Tunacı *et al.* (1999) [40] described CT features of PAA in 12 BD patients, showing that peripheral aneurysms often decreased in size or resolved under medical treatment. In a comprehensive cumulative analysis, Uzun *et al.* (2005) [17] reviewed 598 pulmonary BD cases, confirming that PAA represented the most common thoracic vascular manifestation of the disease. The study also highlighted the strong male predominance (M:F ≈ 6:1) and mean age at onset around 30 years.

More recent cohorts focused on the spectrum of pulmonary artery involvement (PAI). Seyahi *et al.* (2012) [18] analyzed 47 BD patients with PAI, of whom 34 (72%) had aneurysms and the remainder isolated thrombosis. They observed a mortality rate of 26%, mainly related to recurrent hemoptysis. In another Turkish cohort, Yılmaz *et al.* (2021) [41] reported 61 patients with PAI, among whom 11 (18%) presented with PAA, while the majority had isolated pulmonary thrombotic occlusions. Interestingly, intracardiac thrombosis was noted in 20% of cases, indicating extensive vascular inflammation. Geographically, almost all denominator-defined data derive from Turkey, with additional case reports from Iran, Egypt, China, Japan, and Italy. The global compilation by Çelik *et al.* (2015) [7] summarized 207 BD-associated PAA cases reported across 23 countries between 1981 and 2014. Mortality in this dataset approached 30%, despite improvements in therapy. The same review pointed out that Japan—despite its high prevalence of BD—has reported very few PAA cases, suggesting potential ethnic or genetic protection or underrecognition.

Several smaller series from Asia have echoed these findings. Zhang *et al.* (2015) [42] described 15 Chinese patients with pulmonary BD, in whom 9 (60%) exhibited aneurysmal lesions. Naguib *et al.* (2015) [43], in an Egyptian cohort of 15 BD patients, found that 5 (33%) had PAA confirmed by helical CT. Similarly, a multicenter review from Iran noted that vascular involvement affected up to 25% of BD patients, with pulmonary lesions in approximately 2–3% of cases [44].

The introduction of multidetector CT and CT pulmonary angiography (CTPA) revolutionized the detection of small, subsegmental aneurysms. Modern imaging reviews, such as that of Yazgan *et al.* (2021) [45], documented a wide range of thoracic manifestations beyond PAA, including pulmonary artery thrombosis, bronchial occlusions, and infarctions, yet reaffirmed that aneurysms remain the defining vascular lesion. The prevalence of PAA within pulmonary BD cohorts ranges from 10% to 80%, depending on inclusion criteria.

Outcomes have gradually improved. Historical mortality reached nearly 50% in early Turkish series [10], but more recent data suggest that aggressive immunosuppression—particularly with cyclophosphamide and corticosteroids—has reduced fatal hemoptysis [40, 41, 45]. Biologic agents, such as infliximab, have been reported to induce regression of aneurysms in refractory cases [46]. However, long-term survival data remain scarce, and recurrence after apparent remission is not uncommon.

The Hughes–Stovin syndrome (HSS), considered a related but distinct entity, remains exceedingly rare. The systematic review by Khalid and Saleem (2011) [23] identified fewer than 40 confirmed cases in the English literature at the time, while updated estimates through 2024 list under 60 cases globally [26]. The condition typically affects young men and presents with deep vein thrombosis followed by PAA. Clinical and pathological features overlap significantly with vascular BD, supporting the hypothesis that HSS may represent a monosymptomatic or incomplete form of Behçet's disease [24, 25, 47]. No denominator-based epidemiological estimates are available for HSS due to its extreme rarity.

Taken together, the pooled findings from denominator-based cohorts and national series suggest that pulmonary artery aneurysm affects approximately 1% of all patients with Behçet's disease, with regional variation between 0.16% and 1.1%. Mortality remains high—around 30–50% depending on cohort and treatment era—although modern management has improved outcomes. The rarity of HSS and its overlapping phenotype with BD make it impossible to calculate population-based frequencies.

Discussion

This systematic review confirms that pulmonary artery aneurysm (PAA) is an uncommon yet distinctive and life-threatening vascular manifestation of Behçet's disease (BD). Despite the rarity of the lesion, its clinical impact is disproportionate, being one of the main causes of mortality in vascular BD [10, 17, 18, 40]. The pooled denominator-based data indicate a prevalence close to 1%, consistent across independent Turkish cohorts and national registries [7, 10, 18]. When pulmonary BD cohorts and cumulative reviews are considered collectively, the proportion of patients with aneurysms among those with pulmonary involvement can reach up to 70%, confirming that PAA is the dominant form of thoracic vascular disease in BD [17, 41, 45].

The predominance of studies from Turkey reflects both the epidemiological burden of BD in that region and the long-standing interest of Turkish investigators in vascular phenotypes. BD prevalence in Turkey is estimated between 80 and 420 per 100,000, compared with 13 per 100,000 in Japan and fewer than 1 per 100,000 in North America [3, 4, 14, 44, 48]. These geographic differences in baseline BD frequency naturally affect the observed incidence of PAA. The absence

of large population-based registries outside the Silk Road region makes cross-regional comparisons difficult, but the available data suggest a genuine east–west gradient.

Ethnic and genetic factors may also contribute. HLA-B51 positivity, known to confer susceptibility to BD, is prevalent in 55–80% of Turkish and Middle Eastern patients but only 13–20% of those in Western Europe [49]. Some authors have hypothesized that certain HLA backgrounds may influence vascular tropism, although conclusive evidence remains lacking [50]. Environmental factors, including infections or mechanical triggers in pulmonary circulation, have been speculated as possible co-factors [51].

Temporal analysis reveals that the apparent frequency of PAA has remained within the same order of magnitude over the past three decades. However, improvements in imaging have markedly changed disease detection. In the pre-CT era, only central and advanced aneurysms were identified, often presenting with massive hemoptysis and high fatality [10, 11, 12]. With the advent of helical and multidetector CT, smaller and peripheral aneurysms began to be recognized earlier, sometimes incidentally [40, 45]. This shift may have contributed to improved survival through timely immunosuppressive therapy.

Modern management relies primarily on corticosteroids and cytotoxic agents such as cyclophosphamide, which induce partial or complete regression of lesions in many cases [40, 52]. The introduction of biologic therapies—particularly tumor necrosis factor (TNF) inhibitors—has further changed the prognosis. Case series have documented resolution or significant reduction in aneurysm size under infliximab or adalimumab [46, 53, 54]. Nevertheless, recurrence after treatment discontinuation has been reported, emphasizing the need for long-term monitoring [55].

Mortality associated with PAA remains substantial despite therapeutic advances. Early studies reported fatality rates approaching 50%, primarily due to aneurysmal rupture and massive hemoptysis [10, 11]. More recent series suggest a decline to around 25–30%, attributed to earlier diagnosis, improved imaging, and more aggressive immunosuppression [18, 41]. Still, deaths continue to occur, especially among patients with multiple or large aneurysms and in those with concurrent deep venous thrombosis or intracardiac thrombi [41, 56]. The long-term prognosis depends not only on the vascular lesions themselves but also on systemic disease control, adherence to therapy, and access to specialized care.

Marked geographic variation exists in reported PAA frequencies. Turkey and the Middle East remain the epicenters, while East Asia—particularly Japan and Korea—show remarkably few cases despite substantial BD prevalence [7, 44, 57]. This discrepancy could reflect differences in genetic background, environmental exposures, or diagnostic thresholds. Similarly, reports from Europe and North America are sparse, typically describing isolated cases in immigrant or mixed-ethnicity populations [21, 58]. In China and Egypt, where BD incidence is moderate, small institutional series have identified PAA frequencies of 2–3% within vascular BD cohorts [42, 43, 59].

The Hughes–Stovin syndrome (HSS) remains an extremely rare disorder that mimics vascular BD. The overlap between the two conditions is now widely acknowledged. Pathologically, both display vasculitis of the vasa vasorum, perivascular mononuclear infiltration, and a tendency to form pseudoaneurysms [23, 24, 25]. The sequence of thrombophlebitis followed by aneurysm formation in HSS parallels the venous-to-arterial continuum seen in BD [26, 47, 60]. However, while BD is multisystemic with mucocutaneous and ocular features, HSS often remains confined to the vascular system. Because fewer than 60 cases of HSS have been documented, reliable prevalence data are unattainable [26, 61]. Nonetheless, the demographic profile—young males in the second or third decade—closely mirrors that of vascular BD [62].

The limitations of the present synthesis reflect the deficiencies of the source literature. Most studies are retrospective and hospital-based, leading to referral bias. Denominator-defined cohorts are few and geographically concentrated in Turkey, which restricts generalizability. Heterogeneity in diagnostic criteria, imaging methods, and follow-up duration further complicates comparisons. The absence of standardized definitions for PAA (size threshold, number of lobes involved, or active inflammation) also introduces variability [63]. Moreover, publication bias likely favors severe or fatal cases, inflating apparent mortality.

Despite these caveats, the data demonstrate consistent trends: (1) PAA remains rare but defining in vascular BD; (2) the approximate prevalence of 1% has held stable across decades; (3) mortality, although still high, has declined with modern therapy; and (4) HSS probably represents a clinical variant within the same vasculitic continuum.

Future research should prioritize prospective multicenter registries capable of enrolling unselected BD populations and incorporating standardized thoracic imaging protocols. Integration of CT angiography as part of routine evaluation for BD patients with thoracic symptoms or elevated inflammatory markers could improve early detection. Harmonized definitions of vascular involvement, international data sharing, and genetic correlation studies may elucidate why some populations are spared pulmonary aneurysmal disease. Moreover, longitudinal studies evaluating the long-term effects of biologic therapy on aneurysm regression and relapse are urgently needed [54, 64].

Conclusion

Pulmonary artery aneurysm (PAA) remains an uncommon but distinctive vascular manifestation of Behçet's disease (BD). The pooled data indicate that approximately 1% of all BD patients develop PAA, with prevalence estimates ranging from 0.16% to 1.1% across large Turkish and multinational cohorts [7, 10]. Mortality, although historically approaching 50% [10, 11], has declined with modern immunosuppressive and biologic therapies [40, 41, 46]. Geographic heterogeneity persists, with the highest frequencies reported in Turkey and the Middle East, and comparatively few cases documented in East Asia or Western countries [7, 44, 57]. The extreme rarity of HSS precludes reliable epidemiological estimates, yet its demographic, pathological, and clinical overlap with BD suggests it represents a limited variant within the same vasculitic spectrum [23–26, 47, 61]. Future research should focus on prospective, multicenter registries integrating standardized imaging criteria, unified definitions of PAA, and longitudinal follow-up to assess relapse and mortality trends. The creation of international collaborative cohorts will be crucial to clarifying the global burden of PAA in BD and elucidating the factors underlying regional and ethnic disparities.

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