

Crowned dens syndrome

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Abstract

Background: Crowned dens syndrome (CDS) is clinically characterized by acute cervical pain accompanied by peri-odontoid calcifications. Historically considered rare, recent literature and our clinical findings suggest CDS is not uncommon. This study aimed to evaluate the prevalence, etiology, clinical manifestations, imaging features, and treatment outcomes of CDS.

Methods: We retrospectively analyzed clinical data from 50 CT-confirmed CDS patients admitted to Guangxi International Zhuang Medicine Hospital between January 2019 and March 2025. Patients were stratified into symptomatic CDS ($n = 26$) and asymptomatic CDS ($n = 24$) groups based on the presence of acute neck pain and restricted cervical mobility. Collected data included demographics (gender, age, and smoking/alcohol history), comorbidities (hypertension, diabetes), initial clinical departments, and imaging findings. All symptomatic patients were followed to document treatments and symptom recurrence.

Results: Among 50 radiologically confirmed CDS cases, 26 (52.0%) were symptomatic and 24 (48.0%) were asymptomatic. Calcifications were observed in all peri-odontoid regions on imaging. The overall male-to-female ratio was 19:31 (ratio = 0.61). Symptomatic CDS patients showed significantly higher proportions of females ($p < 0.05$) and older age ($p < 0.05$) compared with asymptomatic cases. Age and female gender correlated significantly with symptomatic CDS, while smoking, alcohol use, hypertension, and diabetes showed no significant association. All symptomatic patients achieved substantial symptom relief with nonsteroidal anti-inflammatory drugs or corticosteroid therapy, including recurrent cases.

Conclusions: Current under-recognition of CDS contributes to high rates of misdiagnosis and underdiagnosis, suggesting its true incidence likely exceeds reported data. Computed tomography (CT) imaging provides definitive diagnostic evidence. Early intervention with nonsteroidal anti-inflammatory drugs or corticosteroids is critical for improving outcomes in symptomatic CDS patients.

Abbreviations: CDS = crowned dens syndrome, CPPD = calcium pyrophosphate dihydrate, CRP = C-reactive protein, CT = computed tomography, HU = Hounsfield units, NSAIDs = nonsteroidal anti-inflammatory, TLA = transverse ligament of the atlas.

Keywords: cervical spine, clinical analysis, crowned dens syndrome, imaging

1. Introduction

Crowned dens syndrome (CDS) is characterized by acute cervical pain with concomitant peri-odontoid

calcifications. Historically, underrecognition of CDS has led to frequent clinical oversight, resulting in misdiagnosis and underdiagnosis. This diagnostic gap subjects patients to unnecessary invasive procedures (e.g., lumbar puncture, biopsy), inappropriate therapies (corticosteroids, antibiotics, antivirals), and prolonged hospitalization—increasing both economic burdens and detrimental impacts on physical/mental health.

CDS predominantly affects elderly populations. Current literature primarily comprises isolated case reports, while patients with chronic cervico-occipital discomfort or asymptomatic presentations often evade diagnosis. Clinicians typically prioritize alternative diagnoses such as cervical spondylosis, fasciitis, or atlantoaxial subluxation,^[1] particularly when patients initially present to nonorthopedic departments. Consequently, CDS has been erroneously classified as a “rare disorder.”

However, accelerated population aging predicts increasing CDS prevalence, transforming it from a perceived rarity to a clinically significant entity. Early prevention,

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detection, diagnosis, and standardized management are therefore imperative. This study proposes a novel classification of symptomatic versus asymptomatic CDS. Through retrospective analysis of clinical data, we comprehensively investigate CDS's diagnostic and therapeutic profile, aiming to enhance clinical awareness and establish evidence-based management protocols.

2. Materials and methods

This study was approved by the Ethics Committee of Guangxi International Zhuang Medicine Hospital Affiliated to Guangxi University of Chinese Medicine. Patients diagnosed with CDS via cervical CT imaging between January 2019 and March 2025 were retrospectively identified by searching our institutional Picture Archiving and Communication System using the keywords "crowned dens syndrome."

The inclusion criteria comprised patients with radiologically confirmed peri-odontoid calcifications. Exclusion criteria included a history of cervical spine surgery, trauma, infection, or neoplastic disease. After applying these criteria, 50 patients were included in the final analysis. All patients underwent cervical CT examinations using either a 64-slice multidetector CT scanner (Philips Brilliance 64) or a 256-slice multidetector CT scanner (GE Discovery CT 750 HD). Based on clinical and radiological findings, patients were classified into 2 groups: the symptomatic CDS group ($n = 26$), consisting of patients presenting with occipitocervical pain, restricted cervical mobility, and CT-confirmed peri-odontoid calcifications; and the asymptomatic CDS group ($n = 24$), comprising patients without cervical symptoms but with incidental findings of peri-odontoid calcifications on CT imaging.

Demographic characteristics (sex, age, smoking history, alcohol consumption), comorbidities (hypertension and diabetes mellitus), initial clinical department of presentation, and comprehensive imaging data were collected for all patients. For those in the symptomatic group, detailed treatment courses were reviewed, and recurrence status was assessed through structured telephone or online follow-up.

CT image reconstruction and analysis were performed using dedicated workstations (Philips IntelliSpace Portal; GE Advantage Workstation 4.4). Multiplanar reconstructions (coronal, sagittal) and 3-dimensional reformatting were generated from the raw data to visualize hyperdense lesions in the peri-odontoid ligaments and soft tissues. The spatial relationship between these calcifications and the atlantoaxial complex was carefully evaluated, with attenuation measurements (in Hounsfield units [HU]) obtained from axial images.

Statistical analyses were performed using SPSS version 27.0 (IBM Corp.). Continuous variables with normal distribution were expressed as mean \pm standard deviation and compared using Student *t* test. Categorical variables were presented as frequencies (percentages) and analyzed using the chi-square test. A *p* value < 0.05 was considered statistically significant.

2.1. Consequence

Among the 50 enrolled patients who met the inclusion criteria, 26 (52.0%) were classified as having symptomatic CDS, while 24 (48.0%) comprised the asymptomatic CDS group. The overall male-to-female ratio was 19:31 (0.61:1). A statistically significant predominance of female patients was observed in the symptomatic group compared with the asymptomatic group ($p < 0.05$). The mean age of symptomatic CDS patients was 71.34 years (range, 31–86), which was significantly higher than that of asymptomatic patients, whose mean age was 64.05 years (range, 44–79).

Within the symptomatic CDS group, male patients accounted for 26.92% (7/26) and female patients 73.08% (19/26). In contrast, the asymptomatic group exhibited an equal sex distribution, with both males and females representing 50% (12/24) of the cohort (Table 1). Among patients with symptomatic CDS, the predominant clinical manifestations included severe cervico-occipital pain (100%), cervical stiffness (69.23%), and dizziness (36.36%). No febrile cases were documented. Statistical analysis confirmed that both older age and female sex were significantly associated with the development of symptomatic CDS. However, no significant correlations were identified between symptomatic CDS and smoking, alcohol consumption, hypertension, or diabetes mellitus. Follow-up of symptomatic CDS patients revealed recurrence of symptoms in 9 of 26 cases (34.61%). The time to recurrence ranged from 0.5 to 12 months, with a median interval of 4 months (interquartile range: 2–7 months).

Among the study cohort, the distribution of initial clinical departments was as follows: 19 patients (38.0%) presented primarily to the Neurology Department due to significant cephalo-cervical pain or nonspecific dizziness. The remaining patients were distributed across orthopedics, rheumatology, emergency medicine, and other specialties (including pain management and rehabilitation medicine) (Fig. 1).

CT imaging analysis revealed that calcifications were located anterior to the odontoid process in 56% (28/50) of patients, followed by posterior (24%, 12/50), lateral (14%, 7/50), and circumferential (6%, 3/50) distributions (Figs. 2 and 3). Atlantoaxial joint disorder or

Table 1
Summary of clinical data of the study patients.

Group	The symptomatic CDS	The asymptomatic CDS
Male	7	12
Female	19	12
Average age	71.34 (31–86)	64.05 (44–79)
Patients ≥ 70 years old	65.38% (17/26)	33.33% (8/24)
Smoking	4	5
Alcohol	3	2
Hypertension	8	5
Diabetes mellitus	4	2

CDS = crowned dens syndrome.

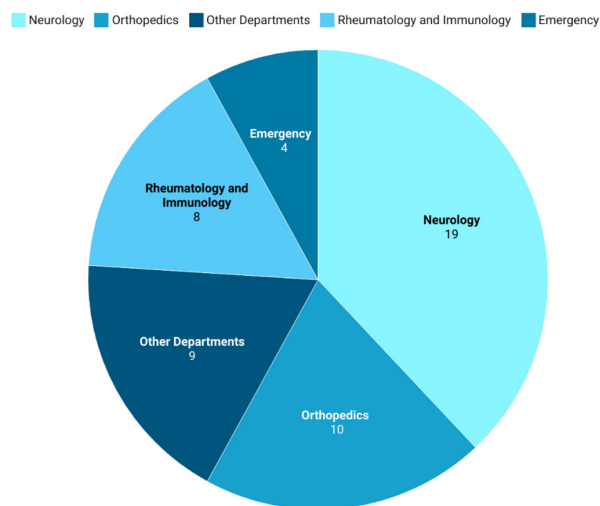


Figure 1. Distribution of initial clinical departments at patient presentation.

subluxation was observed in 18% (9/50) of cases, while 22% (11/50) of patients exhibited stenosis of the atlantal foramen. The mean CT attenuation value of the calcifications measured across all patients was 561.5 ± 115.2 HU.

2.2. Typical case

2.2.1. Case 1. A 63-year-old Asian female presented with acute-onset neck pain and restricted mobility of 1 day's duration, with a visual analog scale (VAS) score

of 4. Her medical history included hypertension and diabetes mellitus, with no history of cervical trauma. Physical examination revealed diffuse tenderness in the occipital and posterior cervical regions, limited cervical range of motion, and exacerbation of symptoms during neck rotation. Spurling's test and foraminal compression test were positive, while the brachial plexus stretch test was negative. Normal physiological reflexes were present, with no pathological reflexes elicited. Cervical CT imaging demonstrated curvilinear calcifications along the posterior and left superior aspects of the odontoid process, forming a characteristic crown-like configuration (Fig. 3). The admission diagnosis was CDS. The patient was treated with nonsteroidal anti-inflammatory drugs (NSAIDs) for symptomatic relief. At 1-week follow-up, she reported complete resolution of pain and restoration of normal mobility. Six months later, the patient experienced recurrence of symptoms with increased intensity (VAS 6). Laboratory investigations revealed significantly elevated C-reactive protein (CRP) (83.56 mg/L) with normal white blood cell count. Repeat cervical CT showed no substantial change in the size or distribution of the atlantoaxial calcifications compared with previous imaging. Based on comprehensive assessment, the patient was diagnosed with recurrent CDS. Another course of NSAID therapy was initiated, resulting in complete symptom resolution within 1 week. Telephone follow-up at 1 year confirmed no further recurrence.

2.2.2. Case 2. A 57-year-old Asian female was admitted to our hospital with a 1-week history of neck pain and restricted mobility. Her symptoms began

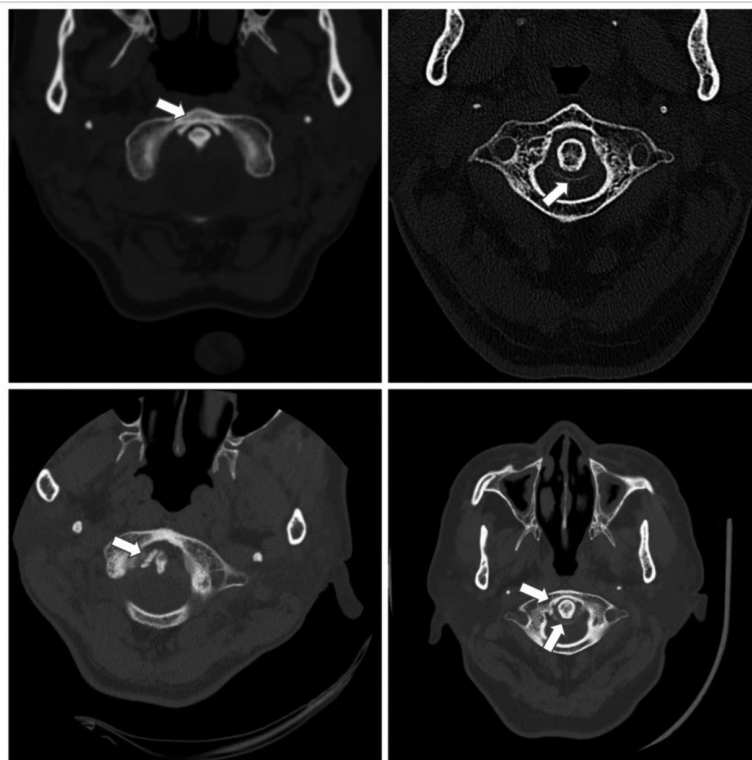


Figure 2. Distribution of calcifications relative to the odontoid process: anterior, posterior, lateral, and circumferential.

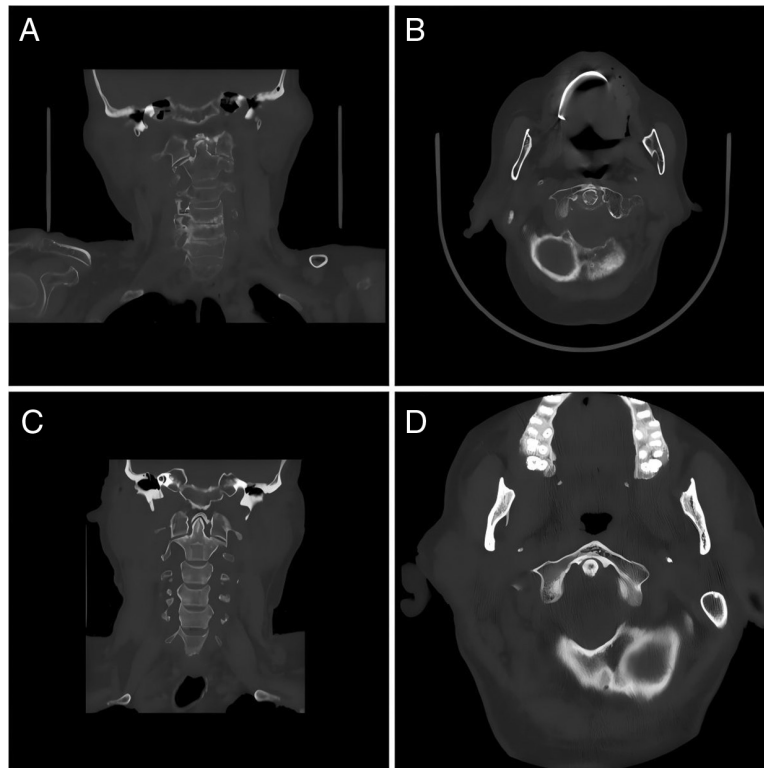


Figure 3. CT images of case 1 and case 2. Panels (A) and (B) display images from case 1, demonstrating calcific deposits located along the posterior and right superior aspects of the odontoid process. Panels (C) and (D) show images from case 2, revealing calcifications distributed at the anterior and superior aspects of the odontoid process. CT = computed tomography.

following a colonoscopy, manifesting as occipital pain and limited rotational movement. Initially diagnosed with muscular torticollis at an external institution, she did not receive any treatment, maintaining a VAS score of 3. Her medical history included gout, with no prior cervical trauma. Physical examination revealed occipital tenderness, restricted cervical motion, and symptom exacerbation during neck rotation. Spurling's test and foraminal compression test were positive, while the brachial plexus stretch test was negative. Normal physiological reflexes were present, with no pathological reflexes elicited. Laboratory investigations demonstrated elevated C-reactive protein (48 mg/L) and erythrocyte sedimentation rate (72 mm/h), with normal serum uric acid levels and white blood cell count. Cervical CT imaging revealed curvilinear calcifications along the anterior and superior aspects of the odontoid process, forming the characteristic crown-like appearance (Fig. 3). The admission diagnosis was CDS. The patient received a comprehensive treatment regimen including cervical traction for 3 days, intravenous ibuprofen (0.4 g, tid), and C2 nerve root block (with compound betamethasone and lidocaine). Within 3 days, her pain and mobility restrictions significantly improved, with follow-up laboratory tests showing normalization of CRP and erythrocyte sedimentation rate levels. The patient was subsequently discharged. Telephone follow-up at 1 year confirmed no recurrence of symptoms.

3. Discussion

CDS, initially reported by Bouvet et al. as acute cervical pain with restricted movement and/or fever,^{1,2} has historically been regarded as a rare entity. Contrary to this perception, our cross-departmental data indicate CDS is substantially underrecognized rather than truly uncommon. Many patients present with subtle or nonspecific symptoms, leading to initial evaluation in nonorthopedic specialties and contributing to diagnostic oversight. In this retrospective analysis, we propose a novel clinical classification distinguishing symptomatic from asymptomatic CDS. Notably, one 31-year-old female with symptomatic CDS developed severe neck pain, potentially precipitated by chronic occupational strain—suggesting CDS may occur in younger populations under specific triggers.

The precise etiology remains elusive, but current evidence implicates microcrystal deposition—predominantly calcium pyrophosphate dihydrate (CPPD)—within the transverse ligament of the atlas (TLA). This incites intense inflammation manifesting as cervico-occipital pain, stiffness, fever, or rarely neuropsychiatric disturbances.¹³ While isolated reports describe hydroxyapatite deposition causing CDS in middle-aged women,¹⁴ CPPD deposition disease—characterized by crystal accumulation in articular cartilage, ligaments, and synovial tissues¹⁵—represents the principal pathophysiology. Although CPPD typically affects peripheral

joints, the craniovertebral junction is a recognized site of involvement. Crystals preferentially deposit in the TLA, tectorial membrane, and atlantoaxial synovium, though mechanistic insights remain limited. Importantly, acute symptoms stem not from mechanical compression but from crystal-induced inflammation: phagocytosed CPPD crystals trigger potent cytokine cascades.^[6] However, this inflammatory response does not invariably elicit a febrile response. In the present series comprising 26 symptomatic CDS patients, all subjects reported intense cervico-occipital pain, with 18 cases (69.2%) demonstrating restricted cervical mobility. Notably, none of the symptomatic patients presented with fever. These observations collectively indicate that fever should not be considered an integral diagnostic criterion for CDS.

In this study, we further analyzed the topographic distribution of calcific deposits relative to the odontoid process. Our findings demonstrate that calcifications may localize to various regions surrounding the odontoid, including anterior, posterior, and lateral aspects, with some cases exhibiting circumferential distribution. However, no direct correlation was observed between these distribution patterns and either disease onset or symptom severity. The mean CT attenuation value of these calcifications measured 561.5 ± 115.2 HU, substantially lower than the typical attenuation of approximately 1000 HU observed in the normal atlantoaxial complex. This notable discrepancy in attenuation values may serve as a supplementary diagnostic indicator for CDS, though further validation is required. Structural abnormalities were identified in

a subset of patients, with 9 cases (18%) demonstrating atlantoaxial joint disorder or subluxation and 11 patients (22%) exhibiting stenosis of the atlantal foramen. These observations suggest a potential association between CDS and structural compromise of the atlantoaxial complex, warranting confirmation through controlled comparative studies. Additionally, our evaluation of spinal cord compression revealed a notable clinoradiological dissociation. While CT findings suggested possible spinal canal compromise in some patients, the majority lacked corresponding clinical manifestations of myelopathy (e.g., limb weakness, gait disturbance, or sphincter dysfunction). Among the limited subset who underwent MRI, images revealed mild spinal cord compression at levels beyond C1–C2, yet none exhibited related neurological deficits (Fig. 4). This suggests that while CDS-related calcifications may radiologically impinge upon the spinal canal, they rarely produce symptomatic compression.

Persistent misperception of CDS as rare underlies frequent diagnostic errors.^[7] Patients presenting acutely with pain and fever are often misdirected to neurology or rheumatology. Even within orthopedics, concurrent degenerative changes on radiographs may obscure CDS diagnosis—a pattern corroborated by our findings. Given its significant prevalence in aging populations, heightened diagnostic vigilance is essential to avoid unnecessary interventions. Classic symptom triads (acute pain, stiffness, fever) risk misdiagnosis as meningitis, potentially prompting unwarranted lumbar puncture.^[8] Noncontrast cervical CT remains the diagnostic gold standard,^[6–9]

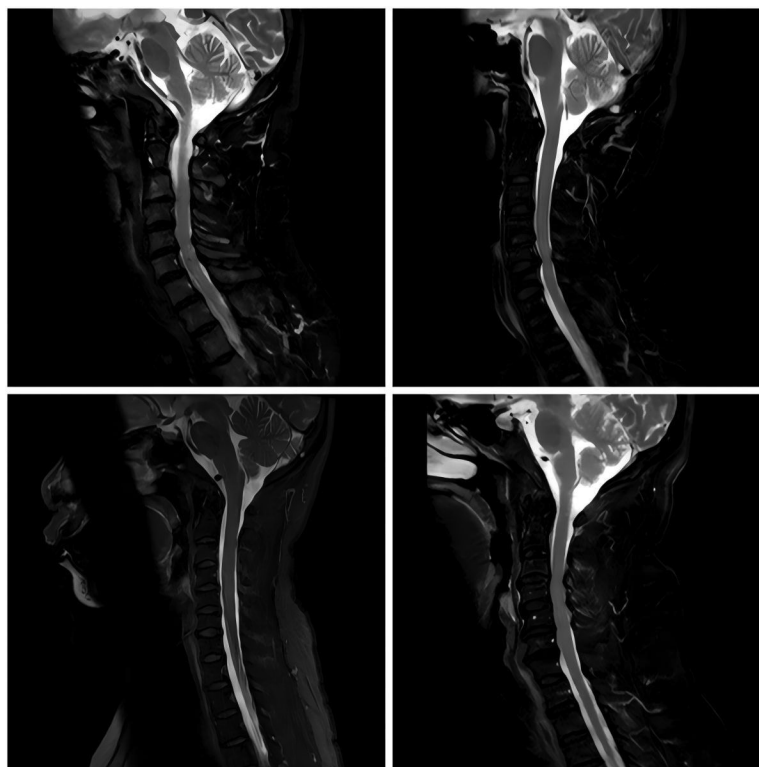


Figure 4. Cervical spine MRI of selected patients. MRI = magnetic resonance imaging.

exquisitely delineating sharp, high-attenuation peri-odontoid calcifications—typically at TLA insertions. While radiographs and MRI may reveal secondary inflammatory changes, CT is indispensable for confirming calcifications and establishing causality between CPPD deposition and acute symptoms. Laboratory studies often show elevated CRP/erythrocyte sedimentation rate but lack specificity; severe symptoms may occur despite minimal CRP elevation or small calcifications.^[9] Synovial fluid analysis demonstrating CPPD crystals provides a definitive diagnosis, but is rarely indicated due to invasiveness. Associations with hypomagnesemia or hyperparathyroidism^[10,11]—potentially via calcium-mediated crystal formation—require further validation.

Therapeutic strategies align with CPPD deposition guidelines: NSAIDs or colchicine constitute first-line therapy, typically yielding symptom resolution within days to weeks,^[5,12,13] while corticosteroids offer effective alternatives. In our cohort, symptomatic patients exhibited robust responses to NSAIDs, further substantiating the CPPD-driven inflammatory basis. However, anti-inflammatory agents do not dissolve established calcifications. Consequently, persistent deposits underlie recurrence—observed in 34.61% (9/26) of our followed cases—despite initial symptom resolution.

This study acknowledges several limitations: the restricted cohort size ($n = 50$) constrains statistical power, while retrospective data collection over extended follow-up periods introduced potential recall bias regarding symptom details. Critically, the absence of posttreatment CT reassessment precludes conclusive analysis of calcification progression and its temporal relationship with clinical recurrence—a significant knowledge gap warranting future investigation.

4. Conclusion

CDS constitutes a distinct clinical entity of CPPD affecting the atlantoaxial junction. The condition arises from selective deposition of CPPD crystals within critical peri-odontoid ligaments—particularly the TLA—forming pathognomonic “crown-like” calcifications. Despite its characteristic presentation, CDS remains profoundly underrecognized in clinical practice, resulting in substantial rates of misdiagnosis and underdiagnosis that likely mask a true prevalence exceeding current literature estimates. Cervical CT serves as the indispensable diagnostic gold standard for definitive evaluation. We therefore advocate prompt utilization of noncontrast cervical CT with 3-dimensional reconstructions for suspected cases to establish an accurate diagnosis. Early therapeutic intervention through immobilization and targeted anti-inflammatory regimens is paramount for symptom control in affected patients.

Acknowledgment

Not applicable.

Ethical statement

We hereby confirm that this study was conducted in accordance with the ethical standards of our institution and the journal's policies. This study involving human participants were reviewed and approved by the Ethics Committee of Guangxi International Zhuang Medicine Hospital Affiliated to Guangxi University of Chinese Medicine (Ethics Approval No. 2024-142-01). The participants provided informed consent to participate in this study. All patient identifiers have been removed to protect confidentiality, and the data presented in the manuscript have been anonymized to ensure privacy.

Conflicts of interest

The authors have no conflicts of interest to disclose.

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Data availability statement

The datasets generated and analyzed during the current study are not publicly available due to our research center policy, but are available from the corresponding author on reasonable request.

Author contributions

Yang-fei Wei contributed to conceptualization, data analysis, performed the literature search, and writing of the original draft paper, and reviewed the paper; Jun-jun Cao contributed to conceptualization, data collection, performed the literature search, patient care, and reviewed the paper; Ming Shi contributed to conceptualization, performed the literature search, and data collection; Tao Wang contributed to data analysis; Jun-peng Liu and Cheng-wang Yang contributed to data collection.

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