






## ORIGINAL ARTICLE

# Crowned dens syndrome: A case series of 72 patients at eight teaching hospitals in Japan

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## Abstract

**Background:** Crowned dens syndrome (CDS) is characterized by calcification around the odontoid process, accompanied by neck pain. Although CDS is supposedly rare, we regularly diagnose and manage this condition, indicating a perception gap between previous studies and our experience. The purpose of this study was to determine the annual incidence of CDS, time to diagnosis in CDS, as well as the features of CDS.

**Methods:** The study design was a retrospective case series study conducted at eight teaching hospitals in Japan. We identified CDS cases from April 2013–March 2015. CDS was diagnosed when patients had acute onset of neck pain and CT showed calcification around the dens and when other diagnoses were unlikely.

**Results:** Seventy-two CDS cases were identified. Mean annual incidence was  $4.6 \pm 2.3$  cases at each hospital. Among those with available data, 57 of 64 had limited rotation (89.1%). The diagnosis of CDS was made in general internal medicine or the emergency medicine department in 61 cases (84.7%). A total of 62 cases (86.1%) were diagnosed within 1 day of presentation, and the median time from initial presentation

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at the hospital to diagnosis was 0.0 days (25th–75th percentiles, 0.0–1.0). For treatment, NSAIDs were used in 56 cases (77.8%) and acetaminophen in 20 cases (27.8%). **Conclusion:** CDS might be more common than has been reported to date. Time to diagnosis of CDS was within 1 day of visiting a teaching hospital. Cervical motion restriction is common in CDS and may be useful in establishing the diagnosis.

#### KEYWORDS

calcium pyrophosphate deposition disease, crowned dens syndrome, general internal medicine

## 1 | INTRODUCTION

Among the different forms of calcium pyrophosphate deposition (CPPD) disease, those that cause acute arthritis are called acute CPP crystal arthritis or pseudogout. Half of these cases occur in the knee. Acute CPP crystal arthritis developing in the cervical spine is called crowned dens syndrome (CDS).<sup>1</sup> CDS is characterized by calcification around the dental process (dens) of the second cervical vertebra (Figure 1) and neck pain.<sup>2</sup> Although there is a recommendation about diagnosis and management of CPPD disease from EULAR, CDS is not mentioned.<sup>3,4</sup> Almost all prior papers pertaining to CDS have concluded that CDS is rare. These papers also note that CDS is easily misdiagnosed as another condition such as meningitis and should be recognized in differential diagnosis. A case report by Oka et al. summarized 69 cases from previous case reports and reported that in 50% of cases, the initial diagnosis was wrong.<sup>5</sup>

Meanwhile, more than 30 case reports and clinical images have been published since 2010,<sup>6–14</sup> and CDS is frequently mentioned in Japanese medical books. We have the impression that we regularly diagnose and manage CDS, suggesting a perception gap between these previous studies and our clinical experience. The only paper that reports the same notion as ours is that of Goto et al. They collected 40 cases of CDS over a 2.5-year period at a single institution in Japan and concluded that CDS is more common than previously recognized.<sup>15</sup> No other English language papers mention the annual number or frequency of CDS. Also, although CDS is said to be easily misdiagnosed, time to a correct diagnosis is not well-known.

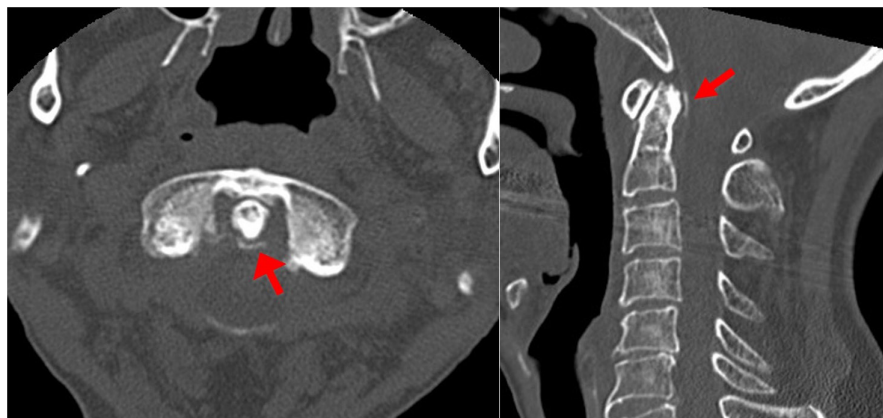
Here, we retrospectively collected data on patients diagnosed with CDS at eight teaching hospitals in Japan over a two-year period. The purpose of this study was to determine the annual incidence of CDS, the number of days required for diagnosis, as well as the features and clinical course of CDS.

## 2 | MATERIALS AND METHODS

A retrospective, multi-institutional case-series study was conducted at eight teaching hospitals in Japan. We enrolled CDS case data from April 2013–March 2015 from these hospitals. CDS was diagnosed when patients had acute onset of neck pain, CT showed calcification around the dens of the second cervical vertebra, and other diagnoses were unlikely.

Data collected from medical records included age, sex, department to which the diagnosing physician belongs, and in- or outpatient onset.

For patients developing neck pain during hospitalization, we also determined the reason for hospitalization; time from outpatient complaint of the symptom to diagnosis; time from the patient coming to the hospital or complaining of the symptom until diagnosis; time from symptom presentation to symptom disappearance; reason for delayed diagnosis; time from initial visit to cervical CT; body temperature; presence of neck pain; limitation of neck flexion, extension, or rotation; difficulty moving; joint pain other than neck pain; disturbed consciousness; posterior cervical tenderness; history of gout or acute CPP arthritis; cervical CT finding of calcification



**FIGURE 1** Left: Axial CT section of the cervical spine at C2. Right: Sagittal CT section of the cervical spine. Both show linear calcification around the dental process of the second cervical vertebra.

around the dental process of the axial vertebra; presence of arthrocentesis, lumbar puncture, cervical MRI, and temporal artery biopsy; findings of cerebrospinal fluid examination; inflammatory indicators such as white blood cell count, C-reactive protein level (CRP), and erythrocyte sedimentation rate (ESR); requirement for hospitalization due to CDS; and treatment with NSAIDs, acetaminophen, glucocorticoids, colchicine, antibiotics, as well as their administration period.

Continuous variables are presented as median (25th–75th percentiles), or mean  $\pm$  standard deviation (SD), and categorical variables as numbers (%).

## 3 | RESULTS

### 3.1 | Incidence

We identified 72 cases of CDS at the eight hospitals over 2 years. Mean incidence was  $4.6 \pm 2.3$  cases per year at each hospital.

### 3.2 | Characteristics

Patient characteristics are presented in Table 1. Because this was a retrospective study, some data could not be collected from all 72 patients, and the number with available data are provided in parentheses. Mean age was  $79.4 \pm 12.6$  years, and 38 patients (52.8%) were women. Mean body temperature was  $37.4 \pm 0.8$  °C, 30 patients ( $n = 65$ , 46.2%) had fever  $>37.5$  °C, and 14 patients had body temperature between 37.1 and 37.5 °C. Among physical findings, 37 patients had limited neck flexion ( $n = 60$ , 61.7%), 38 had limited extension ( $n = 57$ , 66.7%), 57 had restricted rotation ( $n = 64$ , 89.1%), 39 had posterior cervical tenderness ( $n = 55$ , 70.9%), 35 had generalized weakness ( $n = 67$ , 52.2%), 15 had pain in other joints ( $n = 72$ , 20.8%), and six had disturbance of consciousness ( $n = 72$ , 18.3%). A history of gout was reported in two cases, a history of CDS in four cases, and a history of pseudogout other than CDS in eight.

Among those with available data, median white blood cell count ( $n = 62$ ) was  $9.45 \times 10^9/L$  (7.28–11.73), median CRP ( $n = 61$ ) was 8.8 mg/dL (3.3–15.8), and median ESR ( $n = 8$ ) was 74 mm/h (61–107). Arthrocentesis was performed in four cases (5.6%), and detected calcium pyrophosphate in all cases, but the puncture site was unknown. Lumbar puncture was performed in 11 cases (15.3%), showing normal levels for cell count, protein, and glucose in the cerebrospinal fluid. Cervical spine MRI was performed in four cases (5.6%). No patient underwent temporal artery biopsy.

## 4 | DIAGNOSIS

We termed the day of visit to the hospital as day 0. If the patient was hospitalized, then the day the inpatient presented with symptoms suggestive of CDS was day 0. The time from a patient coming to the

hospital to diagnosis or from an inpatient complaining of the symptom to diagnosis was 0 days in 51 cases (70.8%) and 1 day in 11 cases (15.3%) (Table 2). All 72 cases were diagnosed as CDS within 7 days. In summary, 62 cases (86.1%) were diagnosed within 1 day, and the median time was 0.0 days (0.0–1.0).

The specialty of doctors who finally diagnosed CDS was general internal medicine (GIM) or emergency medicine in 61 cases (84.7%), orthopedic surgery in seven cases, cardiology in two cases, and neurosurgery and nephrology in one case each. In 10 cases, diagnosis required more than 2 days from the first presentation. The causes of this longer time to diagnosis included inability of the patient to complain well of symptoms due to old aphasia or impaired consciousness, neck pain developing after the first visit, failure to consider CDS in the differential diagnosis, and attribution of other symptoms as a cause of fever (Table 3). Of 62 outpatient cases, 26 (41.9%) cases required admission, due to generalized weakness in 17 cases, disturbance of consciousness in two cases, and suspicion of another condition requiring detailed examination in seven cases. In 10 cases (13.9%), CDS was diagnosed as a new symptom while the patient was hospitalized for other reasons. The initial reason for admission in these 10 respective cases was alcoholic ketoacidosis, acute heart failure, pneumonia, acute heart failure and pneumonia, COPD, pacemaker implantation due to sick sinus syndrome, acute exacerbation of chronic heart failure, non-ST elevation myocardial infarction, lower extremity cellulitis, and acute cerebral infarction.

### 4.1 | Treatment

NSAIDs were used in 56 cases (77.8%), acetaminophen in 20 cases (27.8%), glucocorticoids in five cases (6.9%), and colchicine in four cases (5.6%). In some cases, two or three drugs were used in combination. Median duration between onset and resolution of CDS symptoms ( $n = 29$ ) was 8.0 days (5.0–11.0).

## 5 | DISCUSSION

We retrospectively identified a total of 72 consecutive cases of CDS in eight general hospitals in Japan over 2 years. CDS might be more common than has been reported to date. Further, in the majority of cases, time to diagnosis of CDS was within approximately 1 day of visiting a teaching hospital. Although CDS has been reported to be rare and often misdiagnosed, we found that delays in diagnosis were limited to certain special circumstances. Additionally, CDS was associated with a high probability of cervical rotation restriction, affecting as many as 89% of cases.

We found that the incidence of CDS was 4.6 cases per year at each hospital. We did not collect data of a control disease to compare with these reported numbers. Among previous studies, Goto et al. conducted a single-center prospective study in Japan and reported 40 CDS cases in 2023 patients with neck pain over 2.5 years.<sup>15</sup> Takahashi et al. reported 35 CDS cases over 4 years from

TABLE 1 Patient characteristics.

Variable	Number of samples	
Age, years	72	79.4 ± 12.6
Sex, Female, n (%)	72	38 (52.8)
During hospitalization, n (%)	72	10 (13.9)
Diuretic used, n (%)	62	8 (12.9)
Time from patient visit to hospital to diagnosis, or from inpatient complaint of symptoms to diagnosis, days	72	0 (0-1)
Body temperature, °C	65	37.4 ± 0.80
Fever more than 37.5 °C, n (%)	65	30 (46.2)
Neck pain, n (%)	72	72 (100)
Difficulty moving, n (%)	67	35 (52.2)
Joint pain other than neck pain, n (%)	72	15 (20.8)
Past history of gout, n (%)	69	2 (2.9)
Past history of acute CPP arthritis other than CDS, n (%)	70	8 (11.4)
Past history of CDS, n (%)	70	4 (5.7)
Disturbance of consciousness, n (%)	72	6 (8.3)
Posterior cervical tenderness, n (%)	55	39 (70.9)
Limitation on neck flexion, n (%)	60	37 (61.7)
Limitation on neck extension, n (%)	57	38 (66.7)
Limitation on neck rotation, n (%)	64	57 (89.1)
Arthrocentesis, n (%)	72	4 (5.6)
Lumber puncture, n (%)	72	11 (15.3)
Mononuclear leukocytes (μl)	11	1 (1-2)
Polymorphonuclear leukocytes (μl)	11	0 (0-0)
Protein (mg/dl)	11	38 (29-44)
Glucose (mg/dl)	11	72 (64-114)
White blood cells (/μl)	62	9450 (7280-11,730)
CRP (mg/dl)	61	8.8 (3.3-15.8)
ESR (mm/hour)	8	74 (61-107)
Cervical MRI, n (%)	72	4 (5.6)
Hospitalization required due to CDS, n (%)	62	26 (41.9)
Treatment		
NSAIDs, n (%)	72	56 (77.8)
Acetaminophen, n (%)	72	20 (27.8)
Glucocorticoid, n (%)	72	5 (6.9)
Colchicine, n (%)	72	4 (5.6)
Symptom presentation to symptom disappearance, days	29	8 (5-11)

Note: Data are expressed as mean ± SD or median (25th–75th percentiles), or n (%). Number of samples means the number of patients for whom data could be collected. Because this was a retrospective study, data could not be collected from all 72 patients.

TABLE 2 Number of days to diagnosis of CDS.

Day	Number of cases	%
0	51	70.8
1	11	15.3
2	3	4.2
3	1	1.4
4	1	1.4
5	2	2.8
6	2	2.8
7	1	1.4
Total	72	100

Note: The day an outpatient visited the hospital was day 0. If a patient was hospitalized, the day the inpatient presented with symptoms suggestive of CDS was day 0. The time from a patient visiting the hospital to diagnosis, or the time from an inpatient complaining of symptoms to diagnosis, is shown. Percentages may not add up to exactly 100% due to rounding.

two hospitals and one clinic in Japan.<sup>16</sup> A prospective study from a 400-bed general hospital in Iran reported 24 CDS cases in 2 years.<sup>17</sup> These previous and our present findings indicate that multiple cases are reported per institution per year.

Moreover, we found that most cases of CDS were correctly diagnosed within 1 day of visit to a teaching hospital in Japan. CDS was diagnosed in 86.1% within 1 day of hospital visit (or onset during hospitalization) and in all cases within 1 week.

Previous reports have stated that CDS is rare and easily misdiagnosed compared to other diseases.<sup>5,18-20</sup> On the contrary, our findings suggest that it may be easily diagnosed. The reason for prompt diagnosis might be easy access to CT scan in Japanese hospitals or to superior diagnostic capabilities in the facilities participating in the study. Oka et al. found that over the period 1985–2014, 56% of cases of CDS reported in the English language literature were from Japan, and Japan has the highest number of CT scan machines per population in the world.<sup>5</sup> In our present study, most cases of CDS were diagnosed by general internal medicine (GIM) or emergency department physicians. All eight participating institutions in this study are teaching hospitals that have both GIM and Emergency departments. These hospitals are rare in Japan, in that the majority of Japanese hospitals have neither department. It is possible that the physicians at the study sites were able to diagnose CDS quickly because they were particularly skilled at diagnosis.

Regarding cases in which the diagnosis of CDS was delayed, 10 cases (13.8%) took more than 2 days (Table 3). CDS can be overlooked and diagnosis can be delayed if the patient is unable to complain well of symptoms due to old aphasia or impaired consciousness, or if there are symptoms other than neck pain, or if symptoms arise during hospitalization. Aouba et al. reported that two patients with systemic symptoms were suspected of having giant cell arteritis with polymyalgia rheumatica, and a temporal artery biopsy was performed.<sup>21</sup> Because these two patients were suspected of having

TABLE 3 Clinical characteristics of 10 CDS cases which required more than 2 days for diagnosis.

Case	Age	Sex	Chief complaint	Situation at onset	Onset during hospitalization to diagnosis, days	Hospital visit to diagnosis, days	Reason diagnosis took more than 2 days
1	71	M	Fever	Hospitalized due to acute ischemic stroke	2		The patient had aphasia, and pain was difficult to assess.
2	82	F	Fever	Hospitalized due to pacemaker implantation	6		The fever preceded and delayed the finding of the cervical pain.
3	88	M	Disturbance of consciousness, fever	Required hospitalization during outpatient visit due to disturbance of consciousness		2	Disturbance of consciousness delayed finding neck pain on examination.
4	64	M	Neck pain	Required hospitalization during outpatient visit due to difficulty moving		3	At first examination, treatment was started for redness of the right leg as cellulitis. Suspected CDS after hospitalization.
5	83	M	Pain in neck, both shoulders, back	Required hospitalization during outpatient visit due to difficulty moving		4	Hospitalized on Friday due to physical difficulty. CT was performed on Monday.
6	74	M	Fever, difficulty moving	Required hospitalization during outpatient visit due to difficulty moving		5	On admission, the CDS did not show up in a differential diagnosis.
7	64	M	Fever, systemic pain	Required hospitalization during outpatient visit due to purulent knee arthritis		6	Diagnosis of pseudogout and purulent arthritis in the knee. After hospitalization, the development of CDS was noted.
8	80	F	Fever, difficulty moving	Required hospitalization during outpatient visit due to difficulty moving		7	Various tests were performed for fever of unknown origin. CDS was suspected shortly after admission.
9	86	M	Neck pain	Follow-up observation at outpatient clinic		2	Although a high CRP was observed on the first day, he was not diagnosed and was referred to GIM 2 days later.
10	35	F	Neck pain	Follow-up observation at outpatient clinic		5	Diagnosed with tension headache at the first consultation. Outpatient visit to multiple departments and last diagnosis.

other diseases requiring various tests, diagnosis of CDS took days. If CDS is listed in the differential diagnosis and a serious disease is also suspected, it is inevitable that it will take a few days first to rule out the serious disease. Nevertheless, delayed diagnosis among our cases was not as common as has been reported, and most cases were found to be easily diagnosed.

In our study, 10 cases of CDS occurred during hospitalization, two of which took more than 2 days to be diagnosed. Previous studies have also reported multiple cases of CDS during hospitalization for dental infection,<sup>9</sup> post brain surgery,<sup>16</sup> post endoscopic retrograde cholangiopancreatography,<sup>22</sup> pneumonia,<sup>23</sup> and after acute cerebral infarction.<sup>24</sup> New symptoms such as fever during hospitalization may be due to flare-ups of the primary disease, side

effects of medication, or complications of invasive procedures such as catheterization, making CDS diagnosis even more difficult than in outpatients.

With regard to diagnosis, we found that CDS is characterized by a high rate of cervical rotation restriction, seen in 89.1% of cases. This rate was higher than the flexion limitation seen in 61.7% and extension limitation in 66.7%. This study did not distinguish between active and passive rotation or active and passive bending.<sup>25</sup> There is a physical finding of meningitis called Jolt accentuation of headache. A positive test consists of accentuation of headache by horizontal rotation of the head at a frequency of two to three times per second. It was initially said to have a sensitivity of nearly 100% for meningitis, but sensitivity and specificity are now both considered low.<sup>26,27</sup>

On the other hand, inability to rotate the neck, or pain even with slow neck rotation, may be a characteristic finding of CDS. In one case report, a case of bacterial meningitis with fever, headache, and neck pain was misdiagnosed as CDS, but the report makes no mention of cervical rotation restriction.<sup>28</sup> If it is found that cervical rotation restriction with neck pain is less frequent in meningitis, a finding of restricted cervical rotation with neck pain may help diagnose CDS and rule out meningitis.

Here, we found that CDS may not be as uncommon as previously thought. If CDS is not rare, it would be desirable to conduct an analytic observational study using a control group with neck pain other than CDS or meningitis in the future. This will allow an understanding of the diagnostic accuracy of the various findings, including the cervical rotation restriction presented here. The significance of properly diagnosing CDS is that it may reduce the likelihood of other diseases. Tests such as lumbar puncture, temporal artery biopsy, and head MRI, as well as antibiotic use, may be reduced.

It is of interest that, although the majority of CDS cases were treated with NSAIDs, acetaminophen was prescribed in as many as 1/4 of cases. Two previous CDS reports from Japan also used acetaminophen for treatment.<sup>9,20</sup> Acetaminophen has become the first-line drug of choice in the management of mild chronic pain in the elderly due to its safety profile compared to NSAIDs.<sup>29</sup> However, it cannot be said in general that they are superior to NSAIDs even for short-term use in the elderly. Moreover, acetaminophen is not among the agents recommended by EULAR for treatment of acute CPP crystal arthritis – namely, NSAIDs, colchicine, or glucocorticoids<sup>3</sup> – and the efficacy of acetaminophen against CDS is unknown. We also collected information on the duration of treatment, but there were too much missing data to permit evaluation. Further studies are needed to determine whether acetaminophen may be a treatment of choice for CDS.

## 5.1 | Limitations

Multiple diagnostic criteria for CDS have been proposed, and the results can vary depending on which is selected. Bouvet et al. first reported CDS and defined it as acute neck pain and calcification in ligaments and structures surrounding the odontoid process caused by crystal deposition.<sup>2</sup> In a previous report, a case of CDS was diagnosed by puncturing the area near the odontoid process, but this is not common due to the invasiveness of the arthrocentesis.<sup>6</sup> In our present study, we diagnosed CDS when patients had acute onset of neck pain, CT showed calcification around the dens of the second cervical vertebra and other diagnoses were unlikely. This is the same finding as that by Goto et al.<sup>15</sup> Other reports have reported that CDS diagnosis is based on the association of clinical, biological, radiological, and therapeutic signs: acute periodic attacks of cervico-occipital stiffness and feverish pains with biological inflammatory syndrome and radiological identification of periodontal calcifications due to microcrystalline deposits on the retro-odontoid ligament and dramatic resolution of symptoms under treatment with NSAIDs or

colchicine.<sup>21</sup> However, some of our present CDS cases do not meet these criteria.

In addition, CDS may not be fully diagnosed. Even when a diagnosis of CDS is not made, symptoms can be relieved by symptomatic treatment of pain with NSAIDs. We confirmed the department to which the doctor who made the diagnosis belongs, but did not collect data on the position of the doctor who made the diagnosis or the number of doctors involved in the diagnosis. It therefore remains possible that our study missed some CDS cases due to failure to diagnose it.

## 6 | CONCLUSIONS

CDS might be more common than has been reported to date. Most cases in teaching hospitals in Japan are diagnosed within 1 day of presentation. CDS has been reported to be rare and often misdiagnosed, but we found here that delayed diagnosis was limited to special circumstances, such as onset during hospitalization. Diagnosis may be delayed to rule out more serious conditions when there is no complaint of neck pain or generalized symptoms, or if symptoms newly develop during hospitalization. Cervical motion restriction is common in CDS and may be useful in establishing the diagnosis.

### AUTHOR CONTRIBUTIONS

HI conceived the ideas and wrote the manuscript. HI, HK, TH, KM, SN, MS, MI, KK, KH, EH, NI, and HK collected the data, and all of the authors reviewed and edited the manuscript.

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### CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest for this article.

### ETHICAL APPROVAL

The study was reviewed and approved by the ethics committees.

### CONSENT FOR RESEARCH

Informed consent was obtained in the form of an opt-out function on the institutional web site.

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