

# Population-based incidence and clinical characteristics of idiopathic intracranial hypertension

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**Objective:** To validate the diagnosis of idiopathic intracranial hypertension (IIH) from the Swedish National Patient Register (NPR) and investigate the incidence of IIH, as well as co-morbidities and medication use in a large Swedish population-based sample.

**Methods:** We searched the NPR to find all patients  $\geq 18$  years old with the ICD-10 diagnosis code (G93.2) for IIH in Stockholm County from Jan 1, 2006, to Dec 31, 2013. All medical records were reviewed to validate the diagnosis and to collect additional information.

**Results:** We included 207 patients with an IIH diagnosis, of which 135 (65%) were correctly diagnosed when validated by charts review. Eighty-three patients had disease onset during the study period. This gave a yearly incidence of 0.65/100 000. Female-to-male ratio was 6.1:1. Females, mean age 31.0 (CI 28.8-33.1), were younger at time of diagnosis compared to males, mean age 42.9 (CI 36.4-49.5),  $P < .001$ . The most common co-morbidities were obesity (92%), hormonal conditions (21%) and recent infections preceding the diagnosis (21%). Prior treatment with tetracycline derivatives were seen in 9%.

**Conclusion:** The incidence of IIH in Stockholm is in the lower range of previously reported rates, possibly due to a lower prevalence of obesity. A substantial proportion of patients (35%) did not fulfill diagnostic criteria. Disease onset occurs at younger age in females. Co-morbidities were mainly associated with diseases affecting hormonal balance or causing inflammatory activation. These findings raise new hypothetical theories regarding mechanisms involved in IIH pathogenesis.

## KEYWORDS

co-morbidities, epidemiology, idiopathic intracranial hypertension, incidence, pseudotumor cerebri, risk factors

## 1 | INTRODUCTION

Idiopathic intracranial hypertension (IIH) is associated with increased intracranial pressure (ICP) of unknown cause.<sup>1</sup> The incidence is highest among obese women of childbearing age.<sup>2,3</sup> Due to pressure-induced degeneration of optic nerve connections, there is a high risk of permanent visual damage.<sup>4</sup> The typical clinical presentation comprises symptoms of headache, visual disturbances, double vision, pulsatile tinnitus and nausea, although patients can present asymptotically with incidental detection of papilledema.

The annual incidence of IIH ranged from 0.9-2.4 per 100 000 individuals in three large-scale studies performed in the USA, Libya and Northern Ireland.<sup>2,3,5</sup> Additional studies from Japan and Italy have found a much lower incidence, 0.03 in Japan<sup>6</sup> and 0.3 in Italy per 100 000 inhabitants, respectively.<sup>7</sup>

Several risk factors and co-morbidities have been reported to be associated with IIH and summarized in a meta-analysis by Chen et al.<sup>8</sup> However, due to rareness of IIH and few large controlled studies the associations found might be spurious. IIH leads to considerable consequences for patients including long-standing headache, risk of visual

deficits including blindness<sup>4</sup> and probably cognitive dysfunction,<sup>9,10</sup> as well as substantial socioeconomic consequences (high healthcare and medication costs, loss of work ability).<sup>11</sup> Further studies of associated risk factors are important to gain insights into disease background and development of better treatment regimes.

The incidence of IIH in a Scandinavian population is not known. The main purpose of this study was to validate the IIH diagnosis code in the Swedish National Patient Register (NPR) and investigate incidence of IIH in the Stockholm region as well as to study co-morbidities and medication use preceding diagnosis to evaluate factors associated with disease onset.

## 2 | METHODS

The study protocol was approved by the local ethical committee in Stockholm. We used data from the NPR to identify all adult patients ( $\geq 18$  years) with a diagnosis of benign intracranial hypertension (the International Classification of Disease (ICD, 10th revision) code G93.2) received in a medical institute in Stockholm County during the period from Jan 1, 2006, to Dec 31, 2013. The NPR comprises data on: personal register number, age, sex, date of admission and date of discharge, hospital, clinic, main and secondary diagnosis, and data of procedures for all patients in specialized outpatient and inpatient hospital care settings in Sweden.<sup>12</sup> All private and public caregivers (except primary care) are required to report data to the NPR and the coverage of registration is very good. In 2006, the number of patient registration numbers missing in the NPR register from contact with health care was only 0.6% and the number of main diagnoses missing in the NPR the same year was 1%.<sup>12</sup> After additional local approval from each Head of Departments, all medical records from the year preceding the IIH diagnosis until the most recent available record were reviewed by a neurologist (AS). Information gathered included year of diagnosis, presence of co-morbidities, medication use at time of diagnosis (current and within the previous year), presenting symptoms, age, sex, weight and investigation results.

The criteria for the diagnosis of IIH were fulfillment of the modified Dandy Criteria<sup>13</sup> (see Table 1). If evaluation showed signs of disorder known to increase intracranial pressure or abnormal CSF composition, the patients were given the diagnosis secondary intracranial hypertension (sIH) and excluded from the IIH cohort. Many of these patients had received the ICD-10 code of G93.2 due to lack of other more suitable codes. Patients who fulfilled the criteria for IIH albeit with known risk factors associated with IIH (such as certain medications and endocrine disorders) were still classified as having IIH, not sIH, as has been proposed in newer criteria by Friedman et al.<sup>14</sup> Patients who did not fulfill the diagnostic criteria completely due to some missing data in records or incomplete workup but where the overall clinical picture was strongly consistent with IIH were classified as probable IIH.

As a quality control, 10% of the records were randomly selected and verified by a second neurologist and headache specialist (INR). If there was discrepancy in diagnoses made by the two neurologists, the

**TABLE 1** Modified Dandy Criteria for idiopathic intracranial hypertension

Modified Dandy Criteria	
1	<b>Symptoms and signs of raised ICP</b> (Headache, nausea, vomiting, papilloedema, pulsatile tinnitus, transient visual obscurations)
2	<b>Normal neurological examination</b> Except for sixth nerve palsy, papilloedema and visual field defects
3	<b>No cause for increased intracranial pressure on MRI</b> (Often CTV or MRV is required to [also] exclude venous sinus thrombosis)
4	<b>Cerebrospinal fluid (CSF) opening pressure &gt;25 cm H<sub>2</sub>O with normal CSF composition</b>
5	<b>No other explanation for increased ICP</b>

ICP, intracranial pressure; MRI, magnetic resonance imaging; CTV, computer tomography with venous sequences; MRV, magnetic resonance venography; CSF, cerebrospinal fluid.

cases were discussed in the research group; the same applied when the primary neurologist (AS) was uncertain.

Frequency tables (using Excel 2007) were created and stratified by sex, age at IIH diagnosis, fulfillment of diagnosis criteria, co-morbidities alongside other more detailed characteristics. STATA 13 was used to evaluate the data. In order to assess whether the incidence changed over time during the study period, the number of new onset cases per year was divided by the total Stockholm County population  $\geq 18$  years old for each year (official statistics available from Statistics Sweden) and multiplied by 100 000. We did the same using official statistics over female population in Stockholm County  $\geq 18$  years and female population 18-45 years of age when calculating these incidences. We calculated the confidence interval for the mean incidence using the variance for the time period 2006-2013. Age differences by sex were calculated using a simple regression model using age as a continuous outcome and sex as the independent variable.

## 3 | RESULTS

In total, 249 patients had received the diagnosis code G93.2 on at least one occasion under 2006-2013 at inpatient or outpatient clinics in Stockholm County. We excluded 39 patients who were younger than 18 years old at time of diagnosis. Furthermore, three patients were excluded because their medical records were not retrievable. Finally our study encompassed 207 patients with a diagnosis of IIH and available medical records (see study inclusion chart, Figure 1). A total of 27 medical records with atypical symptoms, lack of workup, and complex co-morbidities (such as other eye disorder) were discussed in the research group for a consensus decision. Furthermore, of the total 207 records, 21(10%) were randomly selected and re-reviewed in a blinded fashion as quality control. In one case, there was discrepancy on diagnosis interpretation and the case was discussed to obtain a consensus decision.

After review, 135 patients had IIH (112 fulfilled IIH criteria and 23 probable IIH). Among these, 83 patients were incident cases diagnosed

for the first time during the study period (77 fulfilled IIH criteria and six probable IIH).

Seventy-two patients did not fulfill the diagnostic criteria for IIH. Secondary IH was seen in 16 patients (8%). In eight patients, this was due to pleocytosis in cerebrospinal fluid (CSF) indicating a meningitis of unknown origin (despite investigation). Other examples were sinus

thrombosis, thrombosis in shunt system, osteopetrosis and meningeal carcinomatosis. In 28 patients (13%), an incorrect diagnosis code was given, meaning that, for example, the code for IIH (G93.2) was reported, but the correct diagnosis was systemic hypertension (n=7), intracranial hypotension (n=6), hydrocephalus (n=3), or the code had been incorrectly typed (n=4). In addition, 27 patients (13%) presented symptoms initially interpreted as IIH, but the diagnostic workup later on excluded this disorder. One case was excluded because of poor workup and missing data.

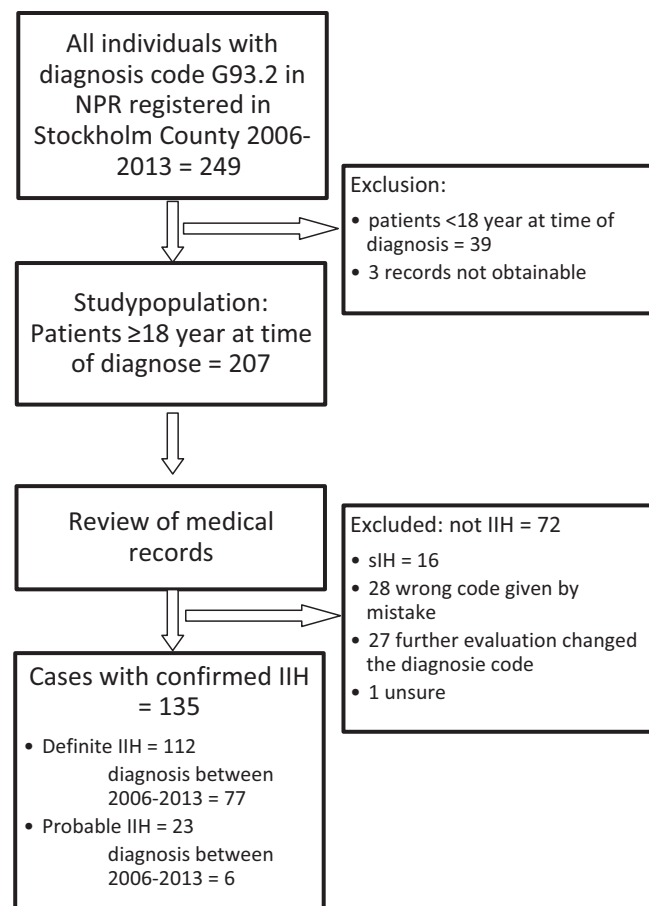
Clinical characteristics (see Table 2) at time of diagnosis in definite IIH patients with sufficient data showed that 84% had symptoms of headache, 72% complained of visual symptoms and 82% had papilledema. Mean opening pressure measured by lumbar puncture (LP) at time of diagnosis was 368 mm H<sub>2</sub>O (SD 94) (n=118). (We did not include patients with probable IIH in these calculations due to frequent lack of information on onset data. Since most of these patients were diagnosed prior to 2003, the initial records were missing).

Most patients (92%) had a body mass index (BMI) over 25 (n=103, including patients with exact BMI value and descriptive terms such as overweight/obese). A precise value was available for 82 patients with mean BMI of 34.7 (SD 0.71). See Table 2.

The average yearly incidence of IIH in the Stockholm County was 0.65 (CI 0.57-0.73) per 100 000 adult inhabitants. Among adult females, the average yearly incidence was 1.1 (CI 0.93-1.28), and when looking at females 18-45 years old, the mean incidence was 1.96 (CI 1.66-2.26) per 100 000 inhabitants (Figure 2).

The female-to-male ratio was 6.1:1. Women were on average younger than men at time of diagnosis; the mean for females was 31.0 (CI 28.8-33.1), and the mean for males was 42.9 (CI 36.4-49.5),  $P < .001$ . Age at onset was missing in four patients.

Co-morbidities reported at the time of diagnosis among IIH patients are presented in Figure 3. Hormonal conditions were present in 29 patients (21%). A recent infection that required medical attention prior to IIH onset was observed in 28 individuals (21%). Hematological disorders were found in seven patients. In males, the most common co-morbidities were hypertension (42%) and hormonal disorders (42%),



**FIGURE 1** Study inclusion chart. NPR, National Patient Register; sIH, secondary intracranial hypertension; IIH, idiopathic intracranial hypertension

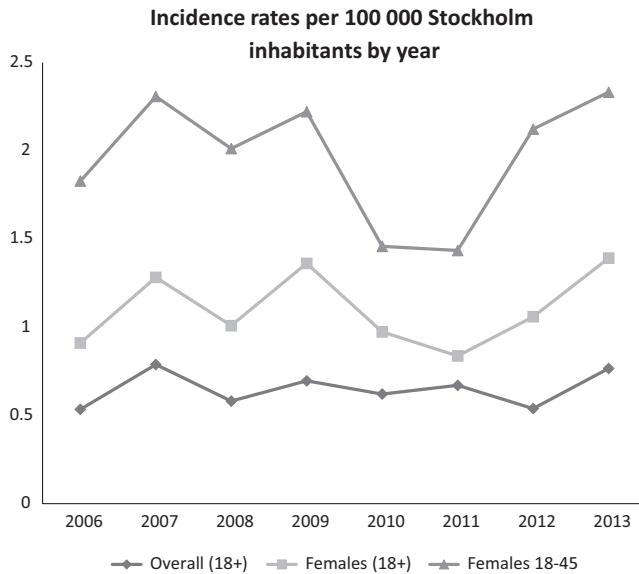
**TABLE 2** Characteristics and symptoms of patients; stratified by definite and probable IIH

IIH symptoms/ characteristics:	Definite IIH (N=112)			Probable IIH (N=23)		
	Yes	No	Data missing	Yes	No	Data missing
Headache symptom	92	18	2	5	5	13
Eye symptom	80	30	2	10	0	13
Papilledema	87	19	6	9	0	14
Mean LP pressure	368 mmH <sub>2</sub> O		4 <sup>a</sup>	311 mmH <sub>2</sub> O		15 <sup>b</sup>
BMI>25 <sup>c</sup>	86	8	18	9	0	14
Mean BMI	34.4 kg/m <sup>2</sup>		34	38.1 kg/m <sup>2</sup>		19

<sup>a</sup>Three lumbar punctures (LP) were done with high values but no precise value declared—for example, >500 mmH<sub>2</sub>O. One LP was not done at diagnosis due to contraindication but done later with an elevated value.

<sup>b</sup>Eleven LPs done prior in older records, in newer records described as elevated but no precise value found. Four LPs not done (one contraindication, one patient refused and two due to extreme obesity).

<sup>c</sup>Including patients with a precise BMI value, as well as those described as overweight/obese.



**FIGURE 2** Yearly incidence of IIH. The yearly incidence of IIH patients over 18 y old in Stockholm County, as well as yearly incidence of females with IIH in female population over 18 y old, and females age 18-45 in Stockholm County

mostly diabetes mellitus (26%). Among males, four out of 19 had obstructive sleep apnea syndrome (21%), but not seen in any females.

Medication use at time of diagnosis is reported in Table 3. Twelve patients had been exposed to antibiotics within the tetracycline family (9%). Treatment with antibiotics and/or antiviral treatment overall was seen in 28 patients (21%). Prior or ongoing corticosteroid treatment was observed in eight patients (6%).

## 4 | DISCUSSION

### 4.1 | Validation of diagnosis

Finding IIH patients using the Swedish National Patient Register (NPR) alone showed a low positive predictive value (PPV) of 65% (definite cases PPV=54%) using the modified Dandy Criteria. Koerner et al.<sup>15</sup> found a similarly low PPV (55%) when validating patient visits at emergency and inpatient clinics, as did Fisayo<sup>16</sup> (PPV 60%) when reevaluating patients at a neuro-ophthalmological department. This indicates that setting a correct diagnosis of IIH can be difficult (particularly early on with frequent change in diagnosis on reevaluation), and coding in registers is also associated with errors (for example, not having a better code to use and typing mistakes when documenting the code).

### 4.2 | Incidence and sex difference

To the best of our knowledge, this is the first report of incidence of IIH in an adult Scandinavian cohort and shows an average yearly incidence of 0.65 per 100 000 individuals. This incidence is somewhat lower than previously reported large-scale epidemiological studies with an incidence between 0.9-2.36 per 100 000.<sup>2,3,5,17</sup> Differences were also present for subgroups such as entire female incidence (1.1

per 100 000) and females aged 18-45 years (1.96 per 100 000) to previous studies where female incidence is reported to be 1.6-4.65 per 100 000<sup>3,5,17</sup> and among females in reproductive age 3.3-12 per 100 000.<sup>2,3,5,17</sup> Our lower incidence may partly reflect the general obesity situation in the population. In Stockholm County, the prevalence of obesity in 2010 was reported to be 10.3% among females and 12.2% among males.<sup>18</sup> This is considerably lower than in Northern Ireland where the reported obesity prevalence in the adult population was 24% in 2013 and IIH incidence was 2.36 per 100 000.<sup>5</sup> A strong correlation between IIH and obesity is known.<sup>8,19</sup>

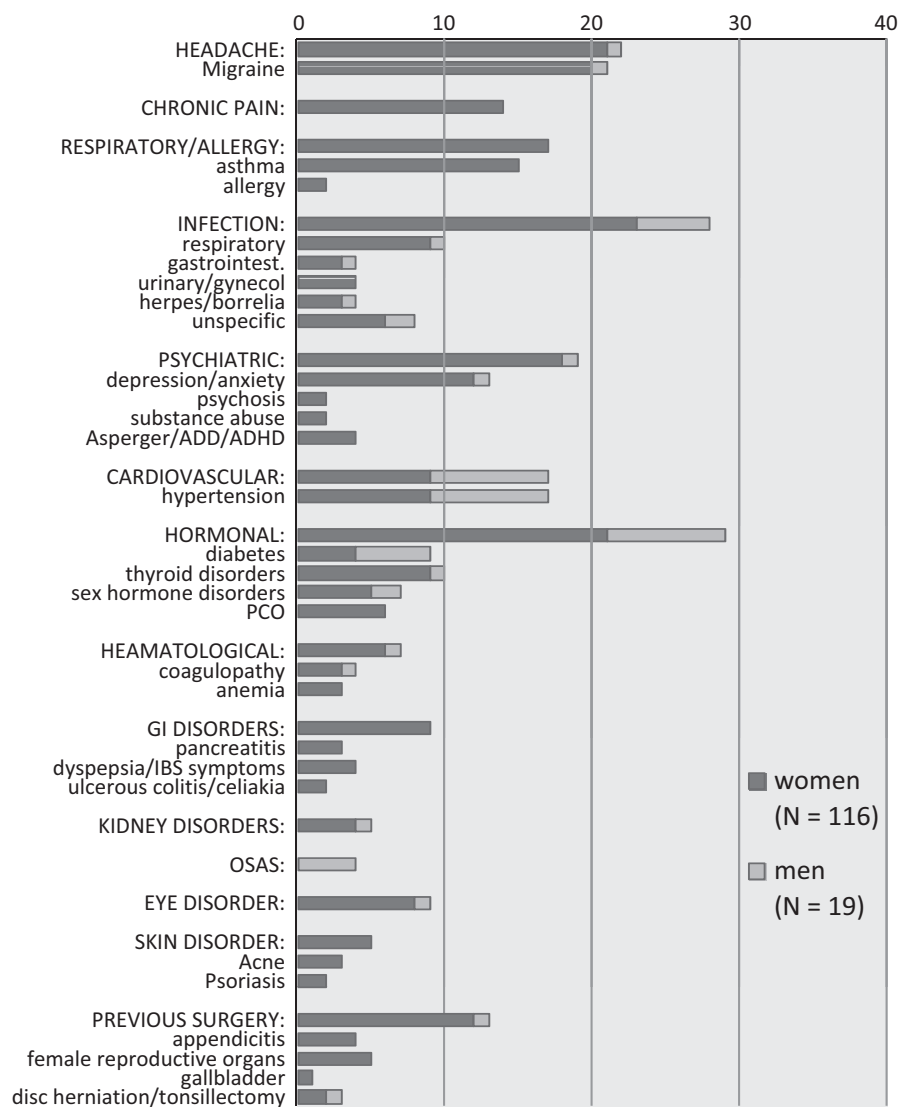
The observed lower incidence of the disease among men in our cohort of IIH patients (14%) is consistent with previous reports.<sup>20,21</sup> Females were younger at disease onset compared to men. This age difference was also reported by Rosenblatt et al. in all their four cohorts,<sup>22</sup> but only reported as a trend<sup>17,20</sup> or not seen in earlier studies.<sup>23,24</sup> Our results indicate a significant sex difference in age at onset of IIH. These results could indicate a sex difference in cause or risk profile behind the disease onset. However, a delay in diagnosing IIH in males could be a possible confounder. Since the disorder is more common in young, obese females this might make doctors neglect thinking of this diagnosis in males with long-standing chronic headache. However, we cannot rule out that chance caused this observed difference.

### 4.3 | Co-morbidities—association with hormonal changes and chronic inflammation in IIH

The most common comorbidity within the IIH population was obesity, with over 90% of patients presenting with a BMI above 25. This is consistent with results from previous studies.<sup>8,19</sup> It is still unknown how obesity is related to IIH. It is increasingly recognized that fat tissue is a source of inflammatory mediators resulting in a low-grade systemic inflammation. Sinclair et al.<sup>25</sup> suggested that the multifactorial pathogenesis of IIH includes dysregulation of glucocorticoid balance and inflammation, a notion that is supported by our findings. The observed frequent hormonal disturbances (21%) and infections (21%) in our study population also support this speculation. Interestingly, a range of conditions also known to be associated with a pro-inflammatory state were found among our IIH patients, such as asthma and allergy, pancreatitis, inflammatory bowel disorders, systemic lupus erythematosus and psoriasis. We speculate that an infection by causing a systemic activation of the immune system with release of pro-inflammatory mediators could act as a triggering factor in IIH development. When looking at our small cohort of sIH, eight patients had developed high intracranial pressure after an infection/inflammatory CNS episode; the majority of these patients showed a prolonged clinical picture similar to IIH patients. This phenomenon has been suggested especially with pediatric IIH cases after viral infections involving the nervous system.<sup>26-28</sup>

Obstructive sleep apnea syndrome (OSAS) was relatively common in males (21%) but not seen in any of the females. Although our male cohort is small, this is in line with the reports from Bruce et al. where OSAS was observed in 24% of the IIH males (total number of males in this study was 66), compared to 4% of the female IIH patients.<sup>18</sup>

## Comorbidities reported in IIH cohort (N = 135)



**FIGURE 3** Co-morbidities reported at time of diagnosis in IIH women (n=116) and men (n=19). n = number of patients. The diagram shows the number of IIH patients with a certain disorder in lowercase letters and organized in groups in uppercase letters. OSAS =obstructive sleep apnea syndrome

**TABLE 3** Medication use possibly related to IIH

Medication	Number of IIH patients	%
Tetracyclines	12	9
Penicillin V	6	4
Other antibiotic <sup>a</sup>	12	9
Contraceptive pill/sex hormone treatment	15	11
Corticosteroid treatment	8	6
Antidepressive drugs	17	13
Levothyroxine	6	4
Lithium	1	1
Indometacin	1	1

<sup>a</sup>Antibiotics not specified in medical records (described as recent treatment of pneumonia, gastrointestinal or urinary tract infection), or other specified antibiotic such as ciprofloxacin, Tazocin, cefadroxil, Selexid. Two patients had been exposed to more than one group of antibiotics.

This sex difference could, however, simply reflect that OSAS is more common in males and prevalence increases with age, or might reflect a possible different pathophysiological mechanism.

In our cohort, 9% of patients developed IIH after treatment with tetracyclines, which has previously been indicated as a likely risk factor for development of IIH. However, as discussed above, an infection/increased inflammation state in itself might also contribute to the triggering of IIH.

#### 4.4 | Methodological considerations

There are limitations to this study. The study population contains some missing data; some data were impossible to find due to diagnostic workup done a long time ago and not transferred to newer medical records; furthermore, some patients had incomplete workup. This may have given an underestimate of the incidence. The reported incidence may also be an underestimation because patients who have not been coded with an IIH code but fulfill the criteria. For example,



those diagnosed only with code for papilledema have not been investigated in our study. We know that ophthalmologists often only put papilledema as diagnosis code on this patient group. Therefore, IIH patients who go for regular checkups regarding their IIH only to ophthalmologists would fall out of our cohort, an issue that would affect prevalence calculation of IIH as we only includes diagnosis code for IIH. However, ophthalmologists always refer the patients to neurologists for evaluation at start of a newly diagnosed papilledema of unknown origin. Hence prevalence has not calculated, but we believe that incidence data would be less affected by this. We also believe IIH is under diagnosed in many cases with chronic headache where evaluation of eyes and lumbar puncture is not performed.

The strength of this study is the review of all medical records to validate all diagnosis codes in the NPR according to the modified Dandy Criteria. The advantage of using this approach is the register's extensive coverage of hospitals and clinics in the Stockholm County, not only the ones in obvious clinical contact with IIH patients. Even though the registry does not cover primary health contacts, we believe this does not affect the validity of this study since IIH is not diagnosed or treated in primary healthcare settings. The co-morbidities were collected by reviewing the records, avoiding possible coding errors. New criteria have been proposed by Friedman et al.<sup>14,29</sup> where all cases with a known possible causation behind the onset of IIH (such as tetracyclines, vitamin A but also including less evident relationships such as anemia) are diagnosed as sIIH. We have chosen not to use these criteria since they are still under discussion and the purpose of this study was to describe co-morbidities as well as pharmacological treatments associated with IIH, since evidence are weak for some proposed associations.<sup>8</sup> Using the older criteria also enables a direct comparison with previous studies showing in fact a low incidence in the Stockholm region.

## 5 | CONCLUSION

We found that IIH is often over- or misdiagnosed and that incidence of this disease in Stockholm is lower than that in other studies, which might be due to lower prevalence of obesity in Stockholm. The study also showed a significant gender difference in age at onset with females being younger. Many of the IIH co-morbidities found in our study are characterized by a pro-inflammatory state in the body, a factor that might be of importance in IIH development.

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## CONFLICT OF INTEREST

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