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The epidemiology of Budd–Chiari syndrome in France

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ABSTRACT

Introduction: Epidemiological data is lacking on primary Budd–Chiari syndrome (BCS) in France.
Methods: Two approaches were used:
(1) A nationwide survey in specialized liver units for French adults.
(2) A query of the French database of discharge diagnoses screening to identify incident cases in adults. BCS associated with cancer, alcoholic/viral cirrhosis, or occurring after liver transplantation were classified as secondary.
Results: Approach (1) 178 primary BCS were identified (prevalence 4.04 per million inhabitants (pmi)), of which 30 were incident (incidence 0.68 pmi). Mean age was 40 ± 14 yrs. Risk factors included myeloproliferative neoplasms (MPN) (48%), oral contraceptives (35%) and factor V Leiden (16%). None were identified in 21% of patients, ≥2 risk factors in 25%. BMI was higher in the group without any risk factor (25.7 kg/m^2 vs 23.7 kg/m^2, $p < 0.001$).
Approach (2) 110 incident primary BCS were admitted to French hospitals (incidence 2.17 pmi). MPN was less common (30%) and inflammatory local factors predominated (39%).
Conclusion: The entity of primary BCS as recorded in French liver units is 3 times less common than the entity recorded as nonmalignant hepatic vein obstruction in the hospital discharge database. The former entity is mostly related to MPN whereas the latter with abdominal inflammatory diseases.

1. Introduction

Budd–Chiari syndrome (BCS) is defined as an obstruction of the hepatic venous outflow tract located anywhere from the small hepatic venules to the entrance of the inferior vena cava into the right atrium, with the exclusion of cardiac and pericardial diseases, and sinusoidal obstruction syndrome/veno-occlusive disease [1]. Primary BCS is characterized by obstruction due to thrombosis or phlebitis, which usually excludes malignant invasion or prior cirrhosis. The available estimates of the incidence and prevalence of primary BCS in European countries are based on either small samples of individual cases collected from hospital liver units over a long period or from large queries of national databases of hospital discharge diagnoses [2–5]. Recent estimates differ widely depending on the studies, which are also markedly different. In France, an unpublished survey performed in academic liver units recorded 20 new cases of primary BCS in the adult population, corresponding to an estimated incidence of 0.36 per million inhabitants (pmi) in 1990 [5]. On the other hand, the estimated incidence in a recent population based study of hospital discharge diagnoses in Northern Italy was 6 times greater, with 2.1 pmi per year between 2000 and 2012 [4].

Because of these limited and conflicting data, the aim of the present study was to estimate the incidence, the prevalence and the main characteristics of primary BCS in French adult residents based on two different approaches: a nationwide questionnaire in specialized hospital liver units, and a query in the French database of hospital discharge diagnoses.

2. Materials and methods

2.1. Questionnaire

Between January 1st and December 31st, 2010, we performed a declarative survey in all 32 French academic liver units including 14 groups in *Ile de France* (an administrative area that includes 18% of the French population), as well as in 16 large non-academic liver units throughout the country (Supplemental material 1). These liver units function as regional reference centers for vascular liver diseases since the French Ministry of Health created a nationwide network in 2005. We recorded incident BCS and the prevalence of primary BCS in inpatients or outpatients. Patients under 18 years of age who were not French residents, who developed BCS after liver transplantation, and those with malignancies except for myeloproliferative neoplasms (MPN) were excluded. The date of the first imaging test confirming the diagnosis of BCS was considered to be the date of the initial diagnosis. Characteristics at diagnosis were recorded using a standardized questionnaire, including the risk factors for venous thrombosis [1]. Deficiencies in protein C, protein S and anti-thrombin were not considered to be risk factors because of the difficulty of confirming them as primary defects in patients with liver diseases [1,6]. For similar reasons, antiphospholipid syndrome was only considered to be a risk factor if it was diagnosed before BCS or if systemic lupus was present. This observational survey received approval of the *Commission nationale de l'informatique et des libertés* (CNIL) and *Comité de protection des personnes (CPP) – Nord-Ouest France*.

Incidences and prevalences were calculated using the 2010 population census performed by the *Institut National de la Statistique et des Etudes Economiques* (INSEE), for residents of France who were 18 years or older. Incident cases of BCS were defined as cases whose diagnosis was confirmed in 2010.

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