standard deviation (DS) of normal, recurring infections, some patients have autoimmune diseases and tumors, absence of isohemagglutinins and lack of production of antibodies (Ab) after vaccination and have excluded other causes of hypogammaglobulinemia.

Objective: Determine the prevalence of CVID in the Specialty Hospital—Centro Medico Nacional Siglo XXI, IMSS, Mexico City.

Methods: We included all patients with suspected diagnosis of CVID, clinical history, laboratory tests, determination of serum Igs and isohemagglutinins and were given a dose of gamma globulin 500 mg/kg/dose every 21 days. The results were analyzed with descriptive statistics. The protocol was approved by the local research committee.

Results: A total of 15 patients, 11 women and 4 men, mean age 34 years (± 11). 14 patients met all international standards and just only one patient has positive isohemagglutinins. The prevalence of CVID calculated according to the total population that is entitled in this hospital, 1,520, 900, 1: 101, 393. In relation to the number of patients served by our service, the prevalence of 0.053% with a ratio of 1:2.533 right holders.

Conclusions: Our results show differences in relation to the published literature, prevalence in female sex ratio of 3:1; participating patients were diagnosed at the 34 years of average age, just only 2 patients were diagnosed before 20 years of age, unlike some of the international and national reports, we establish the definitive diagnosis according to international standards in 93% of cases evaluated. The prevalence of CVID obtained in our study is 1:101.393, higher than reported in international literature is 1:30.000 to 1:50.000, but similar to that reported in Spain in 1997 and Iran in 2006.

600 Infections in 14 Patients with Common Variable Immunodeficiency, Retrospective Study

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Background: Common variable immunodeficiency is a heterogeneous syndrome of primary antibody production failure. It affects 1 in 10,000 to 50,000 individuals, and is the most frequent primary immunodeficiency producing relevant clinical symptoms in adults and children. The hallmark of this disease is recurrent bacterial infections, usually of the respiratory and gastrointestinal tract. Onset is mainly in children aged 1 to 5 years, adolescents aged 16 to 20 years, and adults (fifth decade).

Methods: We assessed retrospectively recurrent infections in 14 patients with definitive diagnosis of CVID, for a period of 2 months through the review of their medical records.

Results: Ten patients were female (71.4%) and 4 were male (28.5%). The average age was 34 years. The average age of diagnosis of CVID was 27.5 years with an age range from 6 to 60 years. In 9 patients (64%) of the total studied CVID diagnosis was made in adulthood. All patients had a history of respiratory infection process in the following distribution: in 9 patients (64%) found a history of bronchiectasis, in 8 patients (57%) was found rhinosinusitis, and pneumonia; in 5 patients (35%) recurrent or chronic otitis media and one patient was a history of pulmonary tuberculosis. The lower urinary tract infection was found in 11 patients (78%), chronic diarrhea in 5 patients (35%), osteomyelitis in 1 patient.

Conclusions: Recurrent infections of the respiratory tract specifically low and high and / or gastrointestinal infections should lead to systematic evaluation in which the primary immunodeficiencies are included as CVID.

REFERENCE
1. García JM, Gamboa P, De la Calle A, Hernández MD, Caballero MT. Diagnosis and Management of Immunodeficiencies in Adults by Allergologists J lnvestig Allergol Clin Immunol. 2010;20:185–194.

601 Autoimmune Thrombocytopenic Purpura Associated with Common Variable Immunodeficiency

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Background: Common variable immunodeficiency (CVID) is a condition characterized by antibody deficiency, and therefore susceptible to recurrent pyogenic infections, cancer and autoimmune diseases. It is a heterogeneous syndrome in primary immunodeficiencies and clinically the most important is often diagnosed in adulthood. Autoimmunity occurs in 5% of the general population, in patients with CVID the percentage increased to 20 to 48%, cytopenias being the most common cause of autoimmunity in these patients. Autoimmune thrombocytopenic purpura and autoimmune hemolytic anemia are the most common autoimmune consequences, occurring in 5% to 8% of all patients with CVID. Some patients develop these disorders before the diagnosis of CVID.

Methods: We present the case of a woman of 45 year old, with a history of lower respiratory tract and urinary tract infections in recurrent Pulmonary Tuberculosis. Enter the program short-course treatment strictly supervised for pulmonary tuberculosis with appropriate response. Autoimmune thrombocytopenic purpura refractory to steroids (WWTP) for performing splenectomy.

Results: Anti DNA antibodies, anti nuclear, anti-protease, C. ANCA/PR3 antimioperoxidasa, serology for hepatitis B, C, HIV negative. Serum immunoglobulins were as follow: IgG, 158 mg/dL (normal 700 to 1600), IgM, 55 mg/dL (normal 40–230), IgA, 36 mg/dL (normal 70–400), and, IgE, 38.7 IU/mL (normal 0–100) in more than 2 occasions with values below 2 standard deviations. CD4 T lymphocytes (19%) CD4/CD8 ratio (0.54).

Conclusions: Meets diagnostic criteria for Common Variable Immunodeficiency (CVID) and starting treatment with intravenous immunoglobulin at a dose of 400 mg/kg (every 21 days) with significant clinical improvement and has even managed to integrate into your daily activities. Today, he continues with danazol for WWTP. Therefore, CVID is necessary to consider in the differential diagnosis of autoimmune thrombocytopenic purpura and autoimmune hemolytic anemia in adults (1).

REFERENCE
1. Miguel Park A, et al. Lancet. 2008;372:489–502.

602 Cvid: A Common but Still Underdiagnosed Disease

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Background: Among the more than 150 different forms of Primary Immunodeficiency Diseases (PID) the CVID is the most common symptomat- atic primary immunodeficiency, present mainly in adults. There is a failure of B cells to develop and differentiate into plasma cells; at consequent a reduction of the production of one or more isotypes of antibody can also affected Cell-mediated immunity. Common manifestations included recurrent bacterial infections, that typically involve the upper and lower respiratory tract. Some patients are highly prone to autoimmune manifestations, lymphoid hyperplasia, and tumors.

Methods: We presented 3 cases of CVID with a variety of clinical presentation, evolution and complications related to delayed diagnosis.
Results: A 34 year old male presented chronic diarrhea, weight loss, malnutrition and recurrent upper respiratory infections; digestive tract endoscopy and biopsy was reported with villous atrophy, chronic inflammation and low grade non-Hodgkin’s lymphoma B cell. Unfortunately this patient refused the use of gamma globulin treatment, had a high morbidity, and finally the patient died. The case of a nurse with clinical manifestation of recurrent rinosinusitis and pneumonia, which was diagnosed as IDCV 17 years later, after she developed pulmonary bronchiectasis. Fortunately the disease is under control and she is actually under treatment with intravenous immunoglobulin. Finally, the case of a 44 year old female, who suffered from recurrent upper respiratory infections, additionally had a thyroid gland tumor associated which affecting the thyroid function.

Conclusions: In the 3 cases had low levels of all immunoglobulin as a hallmark. The clinician must be suspecting this condition in all adults with recurrent infectious disease who have gastrointestinal symptoms or who are detected a malignant disease. Early diagnosis and correct treatment are critical in preventing tissue damage, long-term sequelae and death. Replacement with intravenous gamma globulin and antibiotics are the mainstays in the management of these patients.

REFERENCES
1. Shearer W, Fischer A. The last 80 years in primary immunodeficiency: how far have we come, how far need we go? J Allergy Clin Immunol. 2006;117:748–752.
2. Notarangelo L. Primary immunodeficiencies Allergy Clin Immunol. 2010;125:S182–S194.

603 Infections in Patients Diagnosed with Common Variable Immunodeficiency
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Background: Common variable immunodeficiency (CVID) is the most frequent clinically manifested primary immunodeficiency. It is characterized by recurrent infections due to defective immunoglobulin production. We aimed to evaluate the infectious diseases of 7 patients as premising symptoms for diagnosis of CVID.

Methods: All patients had a marked decrease in IgG levels (at least 2 SD below the mean values for their age), a marked decrease in at least one of the isotypes of IgM or IgA, a diagnosis of immunodeficiency at age >2 years, and no other cause of hypogammaglobulinemia.¹

Results: Seven patients who were diagnosed with CVID are investigated for immunodeficiency reasons based on their recurrent infections. Diagnosis of CVID was made at a median patient age of 28 years (range: 16–72 years); of the patients, 6 (86%) were male. All patients were presented with recurrent upper respiratory tract infections (URTI). Additionally, infected bronchiectasis and chronic diarrhea were noted respectively in 3 patients (42.9%), and 2 patients (28.6%); 1 patient (12.3%) had pericarditis. None of them had malignancy.

Conclusions: URTI, pneumonia, and diarrhea are the most frequent initial complications of CVID. CVID often remains misdiagnosed for several years. Unusual length, recurrence, or severity of these infections or pneumonia should suggest the possibility of immunodeficiency and justify appropriate evaluation.

REFERENCE
1. Conley ME, Notarangelo LD, Etzioni A. Diagnostic criteria for primary immunodeficiencies. Representing PAGID (Pan-American Group for Immunodeficiency) and ESID (European Society for Immunodeficiencies) Clin Immunol. 1999;93:190–197.

604 Phenotypic and Functional Analysis of B Cells in Patients with Common Variable Immunodeficiency
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Background: Common variable immunodeficiency (CVID) is a primary antibody deficiency characterized by a decrease in antibody production and low or normal B cell numbers. To elucidate the clinical and immunological heterogeneity of this condition, we studied 13 patients diagnosed with CVID, examined the status of B-cell maturation in patients with CVID by analyzing IgD/CD27 expression, and we analyze the in vitro B cell differentiation to plasma cells.

Methods: T, B and NK cell populations was analyzed by flow cytometry, expression of CD27 marker was determined to define B cell subsets; we also assessed molecules important for B cell proliferation and differentiation, such as TNFRSF13B (TACI), inducible costimulator (ICOS), CD154, CD20, ICOSL and BAFFR. For B cell differentiation assays, total PBMCs were cultured with CpG alone, or with SAC Cowan, Pokweed and CpG; flow cytometric analysis of plasmablast generation was performed after 7 days of culture.

Results: Reduced numbers of T and B cells was observed in CVID patients, this reduction was more prominent in adults than in children. One group of 8 patients showed a significant reduction in IgD+/CD27+ memory B cells while 3 patients had similar percentage than the healthy control group. The IgD-CD27+ memory B cell population was low in 10 patients (<12%); while it was similar to the healthy control group in 2 of the patients. BAFFR expression in B cells was reduced in 4 patients. Finally, the differentiation to plasmablasts was reduced in patients, stimulation with CpG induced 18.5% of plasmablasts (SD = 12.5%) whereas it was 24% (SD = 8.3%) in healthy controls.

Conclusions: These results suggest that a combined defect in T and B cells may account for CVID, at least in some patients. On the other hand, the complete analysis of markers important for B cell proliferation and differentiation such as ICOS, CD40, CD154 and TACI can be a useful tool for understanding this heterogeneous disease. B cells from CVID patients fail to progress to IgD-CD27+ memory B cells and plasmablasts. Based on these facts, we hypothesize that one or more crucial signaling molecules is required to induce terminal differentiation into memory B cells, if defective, may cause CVID.

605 X-linked Agammaglobulinemia: Report of 4 Cases of Mexican Patients at Civil Hospital of Guadalajara Dr Juan I. Menchaca
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