**Supplementary material**

**Malignancy in Common Variable Immunodeficiency: Systematic Review and Meta-Analysis.**

**Kiaee et al.**



**Figure S1**: Meta-analysis of the prevalence of malignancy in CVID patients during the years. Size of circles indicated the sample size of each study

Table S1. **Main characteristics of the included studies on the prevalence of malignancy in CVID patients**

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| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Number** | **First author (year of publication)** | **Reference number** | **Implementation year; Country** | **Study design and sampling method/ Recruitment setting** | **Total sample size (M and F)** | **Age characteristics for total sample** | **Sample size for cancer evaluated** | **Number of reported cancers (%)** | **Detail of reported cancers** |
|  | Kralickova, (2019) | (1) | 1997-2016  Czechia | Retrospective clinical and laboratory data of 295 enrolled patients were obtained from medical records of national referral centers for the treatment of adult patients with primary immunodeficiency diseases. The collected data covered the period from 1997 to 2016. | 295 | NR | 295 | 22 (7.5) | 25 malignancies were found in 22 patients  6 Gastric carcinoma+ 4 B-cell NHL + 5 B-cell HL + 10 other cancers |
|  | Slade A, (2018) | (2) | 2001-2017  Australia | We performed a comprehensive cross-sectional clinical analysis  of adult patients with PADs managed under clinical immunology services in Victoria from January 2001 to February 2017. | 116 | Age of onset median = 20 yr  Age of diagnosis median=35 yr | 116 | 20 (17.0) | 14 (12.0%) Solid organ + 6 (5.0%) Hematological cancer |
|  | Pulvirenti, (2018) | (3) | 1993-2017  Italy | Data on adult CVID patients  in three University-based PID referral centers located in Central Italy (Rome), Southern Italy (Naples), and Northern Italy (Padua-Treviso) were prospectively collected from 01/01/2001  to 31/12/2017 and retrospectively collected from 01/01/1993 to 31/12/2000. | 455  (m=220, f=235) | NR | 455 | 116 (25.5) | 132 malignancies were found in 116 patients1 |
|  | Azizi, (2018) | (4) | Iran | This retrospective cohort study was performed on recorded data of all registered patients with PADs who were diagnosed and treated in Research Center for Immunodeficiencies  at the Children’s Medical Center affiliated to Tehran  University of Medical Sciences during 1995-2016. | 461  (m=311, f=150) | Age of onset median = 1 yr  Age of diagnosis median=7 yr | 243 | 14 (5.8) | - |
|  | Desjardins M., (2017) | (5) | 2017; Canada | Recruitment CVID subjects were recruited through the CPRIMES registry at the McGill University Health Centre (MUHC) and at the Centre Hospitalier Universitaire de Québec. | 42 (M=14, F=28) | Age of onset mean = 18.1 yr  Age of diagnosis mean = 32.1 yrs | 41 | 2 (5.0) | 2 Hematological cancer |
|  | Ferastraoaru D., (2017) | (6) | 2010-2015; USA | This was a retrospective study approved by the institutional review board of the Albert Einstein College of Medicine (Bronx, New York). All adult patients, who had total IgE levels measured from January 2010 through November 2015 at the Montefiore Medical Center (Bronx, New York). | 2339 | NR | 101 | 17 (16.8) | In CVID patients with or Without IgE Deficiency (N=76): 9 Lymphoma + 1 Uterine + 1 Melanoma + 3 Breast cancer.  In CVID patients, who did not have IgE levels measured (N=25): there not-specified malignancy. |
|  | Lauren A, (2017) | (7) | 2014-2016; USA | The United States Immunodeficiency Network (USIDNET) was used to identify individuals with CVID (2013-2016). | 688 | Age at the time of data entry into USIDNET ranged from 3 to 91 years old. | 457 | 58 (12.7) | 21 Lymphoma + 37 types of other cancer 2 |
|  | Valizadeh A.,(2017) | (8) | 2017; Iran | A total number of 235 patients whom were diagnosed as CVID at Children’s Medical Center (Pediatrics Center of Excellence affiliated to Tehran University of Medical Sciences, Tehran, Iran) and were registered in national primary immunodeficiency network database were evaluated. | 120 | Age of onset = IQR=2  Age of diagnosis median = IQR=9 | 120 | 10 (8.3) | 10 Lymphoma |
|  | Furudoi A., (2016) | (9) | 2015; France | Splenectomy samples from 17 CVID patients were obtained between 1978 and 2012. Ten patients were registered in the French DEFI cohort. The remaining 7 patients were followed up in the Department of Internal Medicine, Haut-Le´veˆque Hospital and are enrolled in the ALTADIH Cohort | 17 (M=8, F=9) | Age of onset Mean =20.1 yrs  Age of diagnosis Mean =34.9 yrs | 17 Splenectomized CVID Patients | 2 (11.8) | 1 (F) Lymphoma + 1 (F) Squamous  cell carcinoma |
|  | Janssen W.J.M., (2016) | (10) | 2016; (follow up 3-5 yrs)  Netherlands | All patients were treated at a tertiary care center (University Medical Center of Utrecht, The Netherlands). | 55 (M=30, F=25) | NR | 55 | 3 (5.4) | - |
|  | Kutukculer N., (2016) | (11) | 2014; Turkey | Patients admitted to Ege University Pediatric  Immunology Department | 20 (M=17, F=3) | Age of onset Mean =7 yrs  Age of diagnosis Mean = 8 yrs | 20 | 2 (10.0) | 2 Lymphoma |
|  | Mayor P.C., (2016) | (12) | 2003-2015; USA | United States Immune Deficiency Network (USIDNET) database. Subjects needed their PIDD diagnosis and age of last follow up to be entered into the database by July 1, 2015. | 3658 | NR | 1285 | 119 (9.3) | 37 Lymphoma and 82 types of other cancer 3 |
|  | Gibert M.P., (2015) | (13) | 2000-2013; Spain | CVID patients diagnosed at  Hospital Sant Joan de Déu between 2000 and 2013. | 27 | NR | 27 | 3 (11.1) | 3 B-cell NHL (2 F with MALToma and 1 M with Burkitt), and one out of three also developed a Low-grade astrocytoma |
|  | Vlkova M., (2015) | (14) | 2014; Czech Republic | All patients were Caucasians of Czech origin and fulfilled the PAGID (Pan-American Group for Immunodeficiency) /ESID (European Society for Immunodeficiencies)  diagnostic criteria for CVID | 26 (M=11, F=15) | NR | 26 | 3 (11.5) | 1 (M) Lymphoma + 1(F) Gastric cancer + 1 (F) Basal cell carcinoma |
|  | Gathmann B, (2014) | (15) | 2004-2012; Europen Society | Patients with a confirmed diagnosis of CVID reported in the European Society for Immunodeficiencies (ESID) Database. | 2212 | NR | 902 | 64(7.1) | 23 Lymphoma+ 41 Solid tumor |
|  | Gouilleux-Gruart V., (2013) | (16) | 2004-2012; France | Patients were included into the French DEFI and the Oxford cohort study | 380(M=168, F=212) | NR | 380 | 14 (3.7) | 14 Lymphoma |
|  | Ramirez-Vargas N., (2014) | (17) | 2012; Mexico | The records of active CVID patients from the Immunology Division of seven different reference centers in Mexico were studied. | 52 | Age of onset median = 13.7 yrs  Age of diagnosis median = 19 yrs | 43(M=23, F=20) | 1 (2.3) | 1 Cutaneous T-cell lymphoma |
|  | Abolhassani H., (2012) | (18) | 2011; Iran | Patients diagnosed with CVID at the Children’s Medical Center Hospital (Pediatrics Center of Excellence in Tehran, Iran) | 93 | NR | 93 | 7 (7.5) | 4 Lymphoma+ 3 Gastric adenocarcinomas |
|  | Bateman E.A.L., (2012) | (19) | 2012; GERMANY | Controls and patient groups were recruited to this study through the Clinical Immunology Department at the John Radcliffe Hospital, Oxford, UK | 58  (M=30, F=28) | 46·4 (17·3–75·2) | 58 | 1(1.7) | 1 Lymphoma |
|  | Resnick E.S., (2012) | (20) | 1986 -2012 New York | Subjects with CVID were seen in the Immune Deficiency Clinic at Mount Sinai Medical Center from 1986 through the present. | 473 (M=208, F=265) | Age of onset median =(F:27 and M:24 yrs)  Age of diagnosis median = (F:33.5 and M:30 yrs) | 473 | 72(15.2) | 39 Lymphomas and 33 types of other cancer 4 |
|  | Lin-Lin W., (2011) | (21) | 2004-2009; China | During the period from January 2004 to December 2009, all patients have been diagnosed with PIDs in the Immunological Division, Department of Pediatrics, Xinhua Hospital, China. | 195 | Age of onset median = 3 yrs)  Age of diagnosis median = 10.5 yrs) | 27 (M=18, F=9) | 4 (14.8) | 1 NHL+3 HL |
|  | Rivoisy C. (2011) | (22) | 2004 -2011; France | The French DEFI study data, from April 2004 to July 2011 | 436 | Age of onset mean =44 yrs  Age of diagnosis median = 32 yrs | 418 | 25 (6.0) | 25 Lymphoma |
|  | Aghamohammadi A., (2010) | (23) | 2010; Iran | The Immunodeficiency Clinic at the Children’s Medical  Center affiliated to the Tehran University of Medical Sciences; From 1984 to 2009 | 93 (M=54, F=39) | Age of onset median = 2 yrs  Age of diagnosis median = 8 yrs | 93 | 10 | 10 Lymphoma (M=5, F=5) |
|  | van de Ven AA., (2010) | (24) | July 1995 and July 2008; Netherlands | Wilhelmina Children's Hospital in Utrecht, The Netherlands | 38 (M=32, F=6) | Age of diagnosis mean = 5.5 yrs | 38 | 1 | - |
|  | Ardeniz O., (2010) | (25) | 2001-2008; turkey | Ege University Medical Faculty Internal Medicine Division of Allergy and Clinical Immunology | 23 (M=13, F=10) | Age of onset median = (F:12.5 and M:15 yrs)  Age of diagnosis median = (F:33 and M:28 yrs) | 23 | 3 | 1 (F) Diffuse B cell lymphoma + 1(M) Diffuse B-cell  lymphoma with Pancreatic carcinoma + 1 (M) Papillary carcinoma of thyroid |
|  | Lucas M., (2010) | (26) | 1980-2007; england | Oxford PID Database (FileMaker Pro version 6, Filemaker Inc, UK) | 132 | NR | 90 | 5 (5.6) | 5 Malignancy |
|  | Malamut G., (2010) | (27) | 1962-2004; France | The medical files of patients with primary hypogammaglobulinemia referred to 10 referral centers in France, between January 1962 and July 2004 | 95 | Age of onset mean = 5.9 ± 4.7 yrs  Age of diagnosis mean = 36.8 yrs | 50 CVID patients with gastrointestinal symptoms | 8 (16.0) | 2 Thymoma + 1 Neurinoma + 1 carcinoma + 1 Cervical carcinoma + 1 Peritoneal carcinomatosis + 2 Lung cancer |
|  | Vajdic C.M., (2010) | (28) | 1990-2008; Australia | The study included Australian children and adults diagnosed with PID, as notified to the Australasian Society of Clinical Immunology and Allergy (ASCIA) PID Registry 1990 to February 1, 2008. | 416 | NR | 416 | 38 | 11 Lymphoma 5 + 1 Leukemia + 2 Gastric cancer + 9 (F) Breast cancer + 2 Thymic cancer |
|  | Yong P.L., (2010) | (29) | 2010; USA | All patients seen at The Children’s Hospital of  Philadelphia who met standard diagnostic criteria  for CVID | 24 (M=14, F=10) | Age of onset =>2 yrs  Age of diagnosis median =84 yrs | 24 | 3 | 1 Lymphoma + 1 Leukemia + 1 Melanoma |
|  | Urschel S.,(2009) | (30) | 1990-2004; Germany | With data collected by the immunology laboratory and  the immunodeficiency clinic, all patients with confirmed or  presumed clinical or laboratory diagnosis of CVID between  1990 and 2004 were identified. | 44 | Age of diagnosis median = 10.4 yrs | 32 | 4 (12.5) | 2 HL+ 1 NHL + 1 Burkitt lymphoma |
|  | Aydogan M., (2008) | (31) | 1992-2005; turkey | The records of 10 patients with a diagnosis of CVID  who were attending the Division of Pediatric Allergy and Immunology at Marmara University Medical Faculty from 1992 to 2005 were studied | 10 (M=6, F=4) | Age of onset median = 4 yrs, Age of diagnosis median = 9.4 yrs | 10 | 4 | 2 NHL+ 1 HL  + 1 Wilms tumor |
|  | Chapel H., (2008) | (32) | 1996-2007; UK | The European Common Variable Immunodeficiency Disorders registry of 7 center | 536 | Age of onset median = 24 yrs  Age of diagnosis median = 33 yrs | 334 | 10 (3.0) | 10 Lymphoid malignancy |
|  | Oksenhendler E.,(2008) | (33) | 2004-2007; France | The French DEFI study data, From April 2004 through April 2007 | 341 | Age of onset median = (19 yrs)  Age of diagnosis median = (33.9 yrs) | 252 | 39 (15.8) | 16 Lymphoma + 1 Kaposi sarcoma + 22 other cancer |
|  | Rezaei N., (2008) | (34) | 2008; Iran | The study population was composed of 24 patients with  CVID who were referred to the Division of Allergy and Clinical Immunology of the Children’s Medical Center Hospital, and 20 age- and sex-matched controls recruited from the medical  personnel of this center and their families | 24 (M=17, F=7) | NR | 24 | 3 | - |
|  | Seve P., (2008) | (35) | 2000-2007; France | The current retrospective study included 18 patients: 4  belonging to an earlier study,  and 14 included in the nationwide French DEF-I cohort of adults with CVID | 18 (M=9, F=9) | Age of onset median = (27.5 yrs)  Age of diagnosis median = (6 yrs) | 18 | 5 | 1 T-cell NHL and 3 B-cell NHL + 1 Corpus uteri cancer |
|  | Ward C., (2008) | (36) | 2008; (follow up: average 10 yrs)  UK | A database is maintained of all patients with an immunodeficiency  disease who are seen and treated by the Department  of Immunology at the Oxford Radcliffe Hospitals | 108 | NR | 108 | 6 | 6 Malignancy  (1 Ovarian cancer +  1 NHL) |
|  | Khodadad A., (2007) | (37) | 1997 – 2004; Iran | In Iran, an Iranian Primary Immunodeficiency Registry has been active since 1997, and 515 cases with a variety of primary immunodeficiency diseases were registered at the  end of 2004. From 1997 to 2004, CVID was diagnosed in 39 patients. | 39 (M=24, F=15) | - | 39 | 2 | 1 HL+  1 Primary lung  lymphoma |
|  | Quinti I., (2007) | (38) | 1999-2005; Italy | In 1999, 26 Italian Centers belonging to the Italian Primary Immunodeficiency Network started to submit data for patients with a CVID diagnosis. | 224 (M=111, F=113) | Age of onset Mean =26.6yrs  Age of diagnosis Mean =8.9 yrs | 12 | 14 | 4 B-cell NHL + 1 Polycythemia + 1 Corpus uteri + 1 Ewing sarcoma + 1 Multiple myeloma + 2 Gastric cancer + 2 Breast cancer + 1 Colon cancer + 1 Rectal cancer |
|  | Ogershok PR.,(2006) | (39) | 1992-2005  West Virginia | A medical record review was performed of children younger than 18 years who presented with CVID between the years of 1992 to 2005 to the West Virginia University immunology clinic after approval of the local institutional review board | 12 | NR | 12 | 1 | 1 Sarcoma |
|  | Tanaka N., (2006) | (40) | 1985-2002; USA | The authors retrospectively reviewed chest radiographs  and chest CT scans of 46 patients with CVID, who underwent either a chest radiograph or a CT examination of the chest between 1985 and 2002. | 46 (M=18, F=28) | Age of diagnosis Mean = 0 days to 29 yrs (5.9 yrs) | 46 | 4 | 4 Lymphoma |
|  | Bates C.A., (2004) | (41) | 1985 -2001; USA | Records of patients with a diagnosis of hypogammaglobulinemia seen between 1985 and 2001 in National Jewish Medical and Research Center | 69 (M=25, F=44) | NR | 69 | 5 (7.2) | 4 B-cell Lymphomas + 1 Gastric cancer |
|  | Piqueras B., (2003) | (42) | 2003; France | All of patients had a well-documented CVID diagnosis according to IUIS criteria | 57 (M=20, F=37) | - | 57 | 5 | 3 Lymphoma (B cell) + 2 Thymoma |
|  | Mellemkjer L.,(2002) | (43) | 1990-2002; Denmark–Sweden | In 1979, all paediatric departments in Denmark (5 million inhabitants)  were asked to report patients known to have primary  immunodeficiency to the Rigshospitalet in Copenhagen | 176 (M=86, F=90) | - | 176 | 16 () | 3 NHL  + 1 HL + 3 Gastric cancer |
|  | Nordoy I., (1998) | (44) | 1998; Norway | included 31 patients with a diagnosis of CVID who attended  the Section of Clinical Immunology and Infectious Diseases, Medical Department. | 34 | NR | 34 | 1 (4.0) | - |
|  | Aukrust, P., (1994) | (45) | 1994; Norway | forty-two patients with primary hypogammaglobulinemia under treatment at the section of clinical immunology and infectious diseases, medical department A, the national hospital, Oslo, were included in the study. | 25 (M=9, F=16) | Age of onset= 2yrs | 25 | 1 () | 1 Lymphoma of lungs |
|  | Herbst E.W., (1994) | (46) | 1993; Germany | Small intestinal biopsy specimen of 10 female and seven male patients with CVID were available for immunohistological  evaluation. | 17 (M=7, F=10) | - | 17 | 1 (5.9) | HL |
|  | Pandolfi F., (1993) | (47) | 1992; Italy | Department of Allergy and Clinical Immunology, La Sapienza University of Rome | 40 (M=19, F=21) | Age of onset mean = 28.5 yr | 40 | 2 (5.0) | 1 Gastric cancer + 1 Thymic cancer |
|  | Kinlen L.J., (1985) | (48) | 1975-1981; UK | Registered data in Medical Research Council (MRC) and patients referred to the immunology departments of Northwick Park Hospital, Great Ormond Street Hospital, or the East Birmingham Hospital in UK. | 231 | NR | 220 | 14 (6.4) | 3 NHL + 11 types of other cancer6 |

1. *Pulvirenti, (2018): 25 Gastric cancer, 38 (33 NHL, 5 HL) Lymphoma , 10 Colorectal cancer, 10 Breast cancer, 6 Thyroid cancer, 5 Large Granular Lymphocytic Leukemia , 4 Lung cancer, 4 Liver cancer, 4 Uterine cancer, body, 3 Uterine cancer, cervical, 3 Prostatic cancer, 3 Pancreatic cancer, 3 other blood cancer (CML, polycythemia vera), 1 Kaposi sarcoma , 13 Other cancer include Bladder cancer, meningioma, melanoma, neuro-endocrine carcinoma, ocular carcinoma, kidney carcinoma, adrenal carcinoma)*
2. *Lauren A, (2017): Hematologic malignancy (leukemia, monoclonal gammopathy of unclear significance, and multiple myeloma), Skin cancer (basal cell carcinoma, squamous cell carcinoma) and Non-hematologic malignancy (breast cancer, cervical carcinoma, ovarian cancer, thyroid cancer, tubular adenoma, unspecified solid tumors).*
3. *Mayor P.C., (2016): 37 Lymphoma , 5 (m=2, f=3) Gastric cancer , 3 (m=2, f=1)Colon , 8 (F) Breast cancer , 5 (m=4, f=1) Thymic cancer , 6 (f=3, m=3) Leukemia , 4 (m=2, f=2) Lung cancer , 23 (m=9, f= 14) Skin cancer,3 (m=3)Bladder, 1 (M)Testicular, 2 Ovarian, 2 (f=2) Uterine,2(f=2) Cervix, 18 (m=6, f= 12) Remaining cancers.*
4. *Resnick E.S., (2012): 39 Lymphoma (23 B-cell NHL , 3 Diffuse large B-cell lymphoma, 4 HL, 5 MALToma , 1 Marginal zone lymphoma/monoclonal B lymphocytosis , 1 Monoclonal B lymphocytosis , 1 Diffuse poorly differentiated lymphoma with IgM-macroglobulinemia , 1 T cell–rich B cell EBV positive lymphoma) , 3 Gastric cancer, 3 Colon cancer , 9 Breast cancer , 3 Melanoma , 2 Lung cancer , 3 Malignancy of unknown primary , 2 Oral cancer , 3 Skin cancer , 1 Hepatic carcinoid tumor , 2 Prostate cancer , 1 Vaginal cancer, 1 Thyroid cancer , 1 Ovarian cancer , 1 Esophageal cancer.*
5. *Vajdic C.M., (2010): 11 NHL include: 1 Diffuse large B-cell lymphoma, 2, Follicular lymphoma, 1 Burkitt lymphoma/leukemia , 1 Small lymphocytic lymphoma ,1 Extranodal marginal zone lymphoma type , 1 Peripheral T-cell lymphoma unclassified , 1 Waldenström macroglobulinemia , and 3 NHL unclassified .*
6. *Kinlen L.J., (1985): 3 Lymphomas , 1 Cervix cancer, 1 Unknown primary cancers, 1 Lung cancer, 1 Breast cancer, 7(3 f, 4 m) Gastric cancer*

*M:Male, F:Female , MALToma:* *Mucosa-Associated Lymphoid Tissue lymphoma, yrs: years, NHL: Non-Hodgkin lymphoma, HL: Hodgkin lymphoma.*

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