مستشفى الملك فيصل التخصصي ومركز الأبحاث
King Faisal Specialist Hospital & Research Centre

Primary Immunodeficiency Disease Registry Accumulative Annual Report 2011-2022

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Annual Report Prepared by the staff of Registry Core Facility BESC

Research Center

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Foreword

Primary immunodeficiency diseases (PID) are a group of heterogeneous disorders resulting mostly from a genetic defect leading to a particular deficiency in the host defense system against infections. In addition to susceptibility to microbial infection, the other non-infectious manifestation is related to disturbed immune regulation. Such dysregulation might cause lymphoproliferative and/or autoimmune manifestations. In 1952 Ogden Bruton described the first case of Agammaglobulinemia. Since then, over 250 forms of PIDS have been described. PIDS are considered to be rare disorders worldwide. Therefore, establishing a database is important to determine the magnitude, and types and spectrum of PID disease encountered in a certain population. Databases worldwide have shown geographical and racial variation in the spectrum of PIDs. Furthermore; consanguineous marriages (first cousin marriages) in Saudi Arabia are high, containing up to 60%. This has provided a background where genetic diseases are abundant in the Saudi population. Population data published on PIDS from Saudi Arabia are limited and does not represent the whole nation. Therefore, the need for ongoing and systematic data collection for PIDS in Saudi Arabia is an important healthcare care strategic planning.

In order to determine the magnitude and spectrum of PIDS in a Saudi Arabia, a PID registry was established at King Faisal Specialist Hospital & Research Center in May 2010. More than a thousand of patients have been registered to date and multiple researchers had used the databased to answer their research questions. Our goal is for this database to be the foundation for a national registry.

Dr.Bander AlSaud

Principal Investigator

Acknowledgment

We would like to thank the following individuals: Dr.Edward Cupler Charmaine of the Research Center for the ongoing support. Dr. Edward Devol chairman of the Biostatistics, Epidemiology, and Scientific Computing Department (BESCD) for the support and continuing collaboration, Samia AlHashim for the statistical analysis, Manal AlMarzoqi and Saleh Alageel for the administrative support, we also like to thank our registrar Nada Bawayan.

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Factor about PIDR

Primary immune deficiency diseases (PIDDs) are rare, genetic disorders that impair the immune system. Without a functional immune response, people with PIDDs may be subject to chronic, debilitating infections, such as Epstein-Barr virus (EBV), which can increase the risk of developing cancer. Some PIDDs can be fatal(1). Registry data from Saudi Arabia are limited to two studies. Both are from a homogeneous population and from only one region of the country, and hence likely does not represent the whole nation.

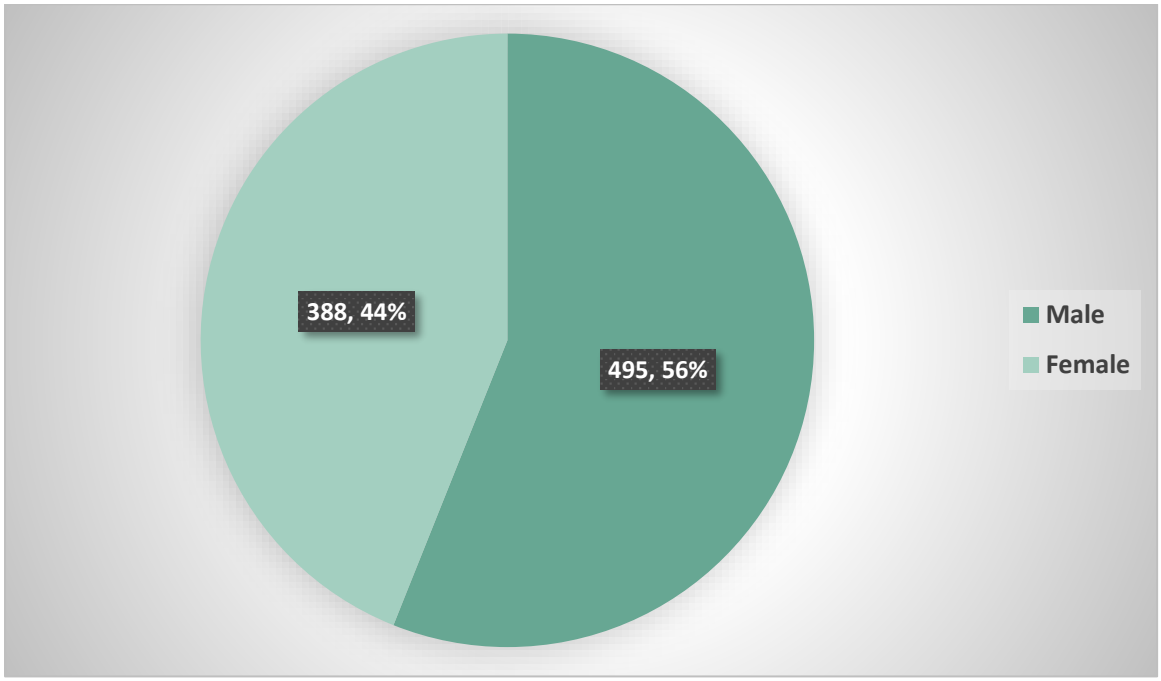
(1) Primary Immune Deficiency Diseases (PIDDs) | NIH: National Institute of Allergy and Infectious Diseases

Primary Immunodeficiency Registry-PIDR

The King Faisal Specialist Hospital and Research Center (KFSHRC) is a tertiary-level hospital for both pediatric and adult patients in the Kingdom of Saudi Arabia. The hospital has received referrals from all around the country since its opening in 1975. A pediatric clinical immunology service was established in 1984 and since then the hospital began to be a major referral for PID patients. In May of 2010, a PID database was established as a prospective ongoing hospital based registry of all patients diagnosed with any of the PID diseases that meet the criteria for diagnosis by the World Health Organization. Data capture forms were developed. Permission was sought and granted by the local Institutional Review Board. The variables identified in the data capture forms are basic demographic, diagnosis details and treatment procedures.

1.0 Demographic Data

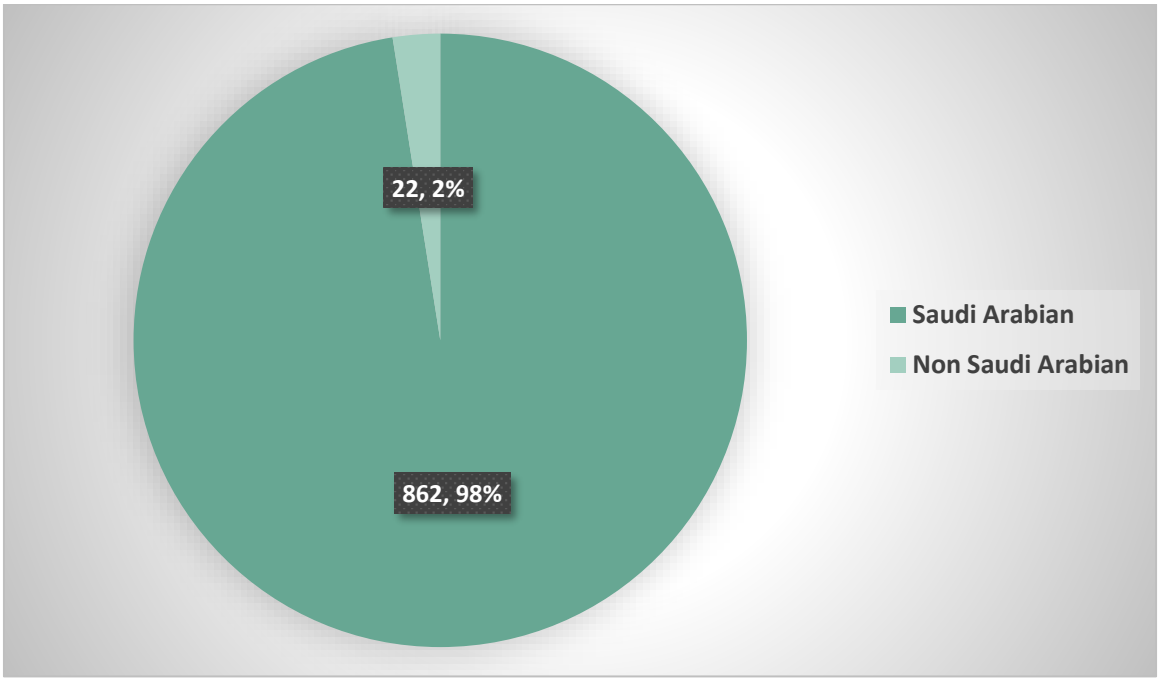
1.1 Gender:



Graph 1.1: Gender distribution in Primary Immune Deficiency patients (n=884).

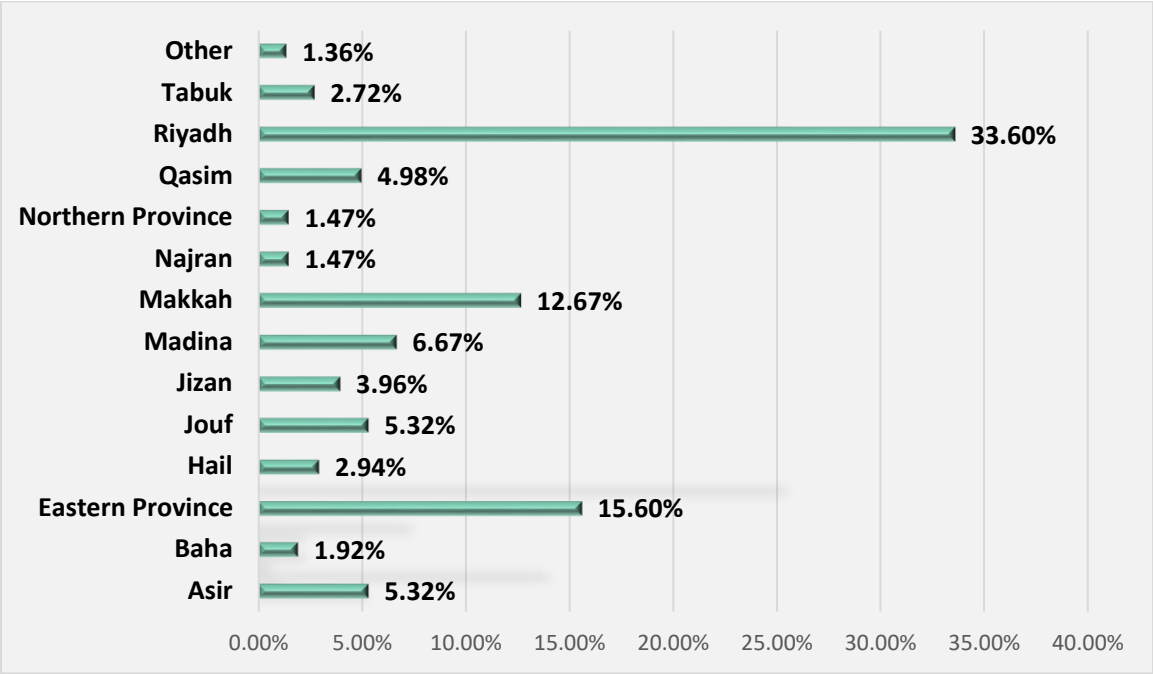
This graph shows that the Males to the Females are almost (1:1).

1.2 Nationality:



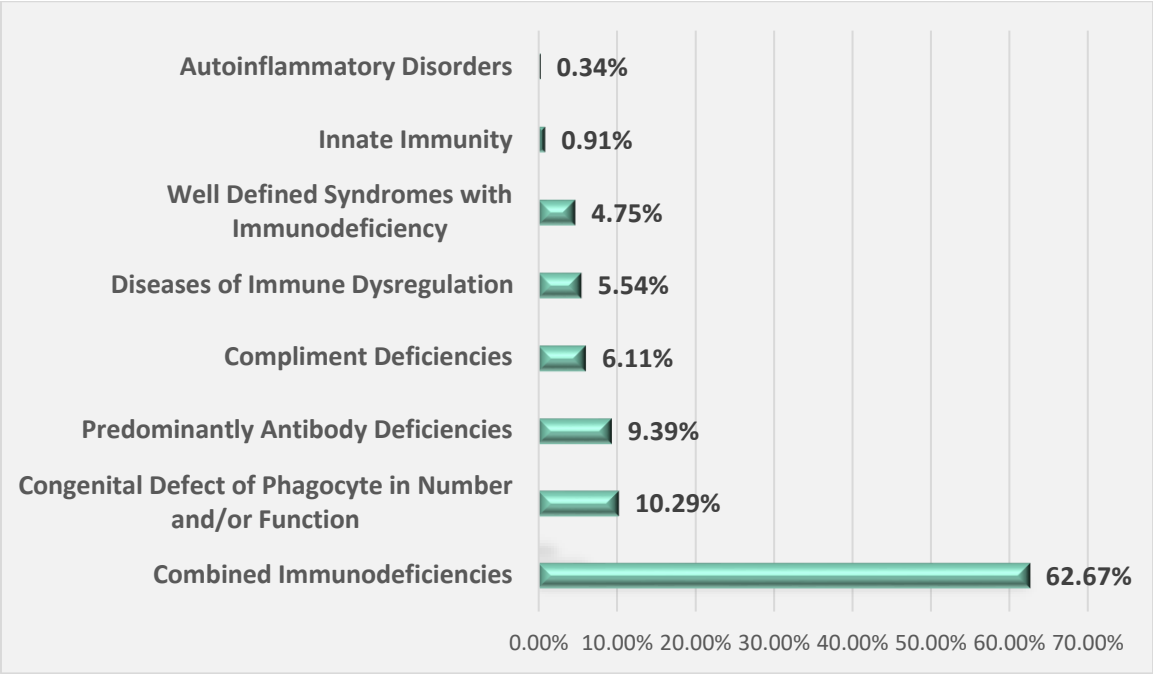
Graph 1.2: Nationality of the Primary Immune Deficiency patients (n=884).
This graph demonstrates that the majority of the patients are Saudi Arabian (98%).

1.3 Current Residence:



Graph 1.3: Primary Immune Deficiency patients current residency (n=884).
This graph shows that the highest place of residence was in Riyadh by (33.60%) and the lowest was in Northern Province and Najran by (1.47%) for each.

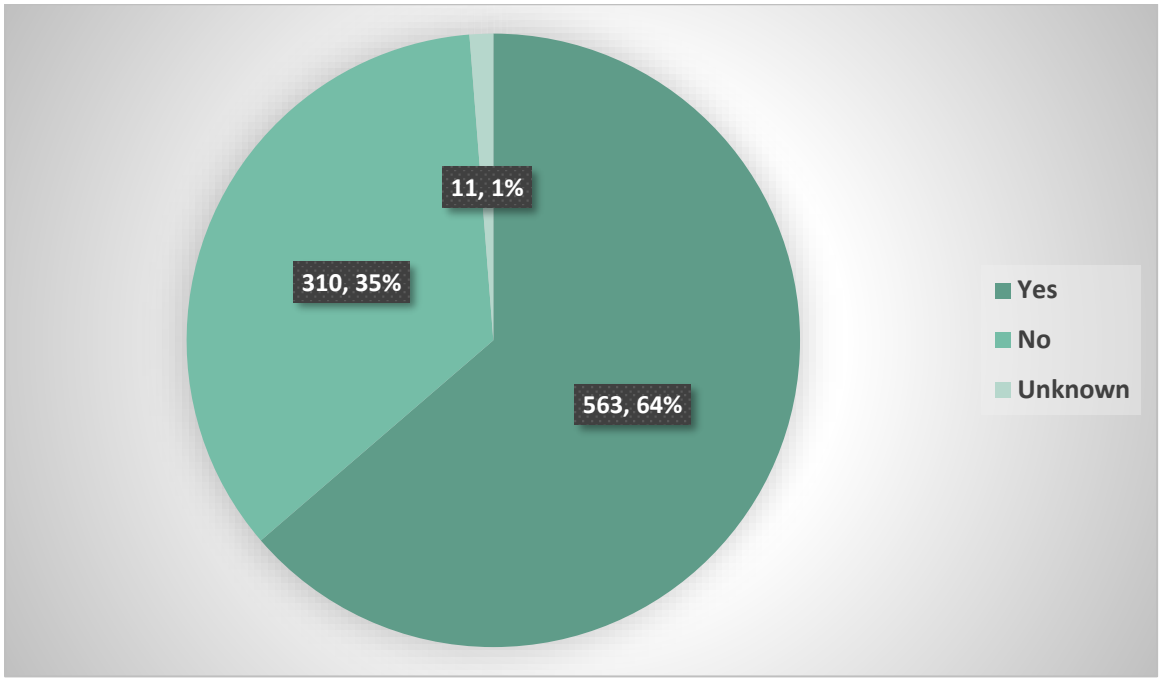
1.4 Diagnosis:



Graph 1.4: Distribution of diagnosis for Primary Immune Deficiency patients (n=884).

This graph demonstrates that the highest diagnosis among the patients was Combined Immunodeficiencies by (62.67%) and the lowest was Autoinflammatory Disorders by (0.34%).

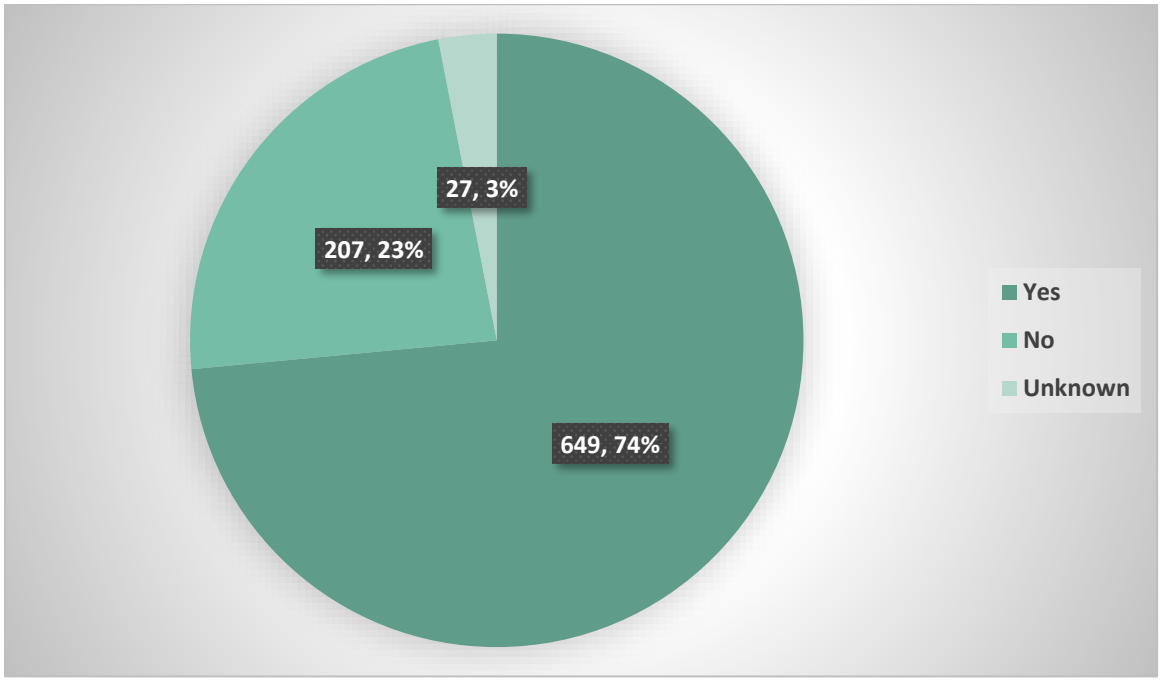
1.5 Family Case:



Graph 1.5: Family cases distribution in Primary Immune Deficiency patients (n=884).

This graph illustrates that PID patients with family case are (64%) and those who are not (35%).

1.6 Consanguinity:

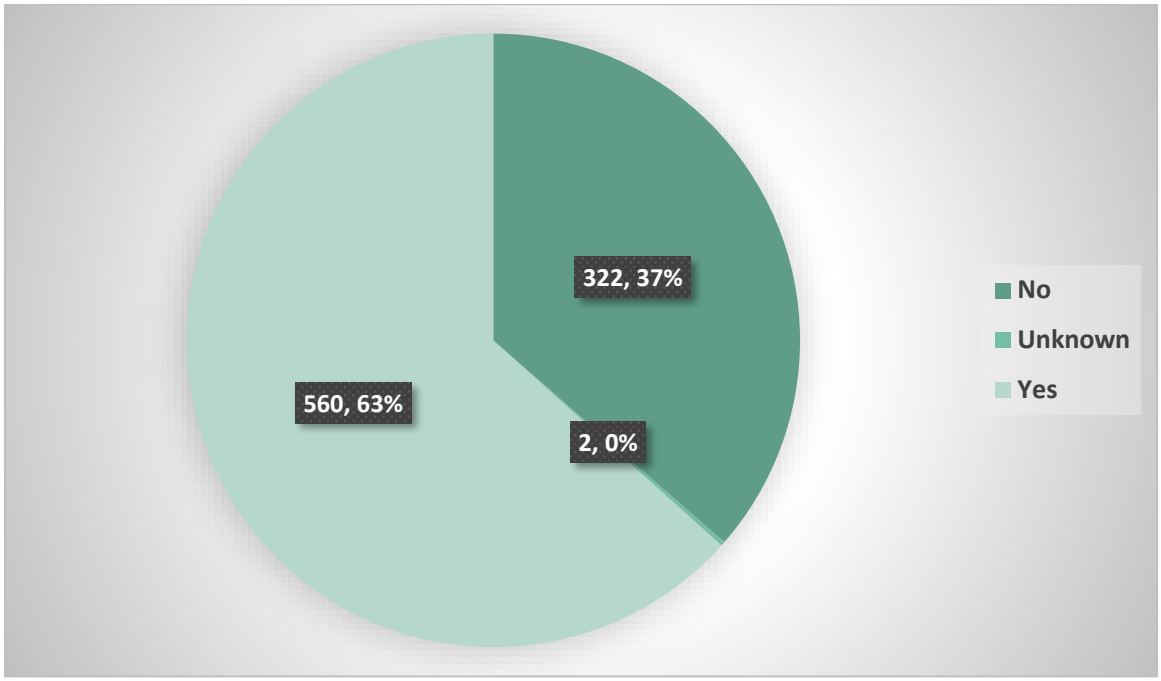


Graph 1.6: Consanguinity distribution in Primary Immune Deficiency patients (n=884).

This graph shows that patients with PID and have consanguinity are (74%) and those who are not (23%).

2.0 Clinical Presentation

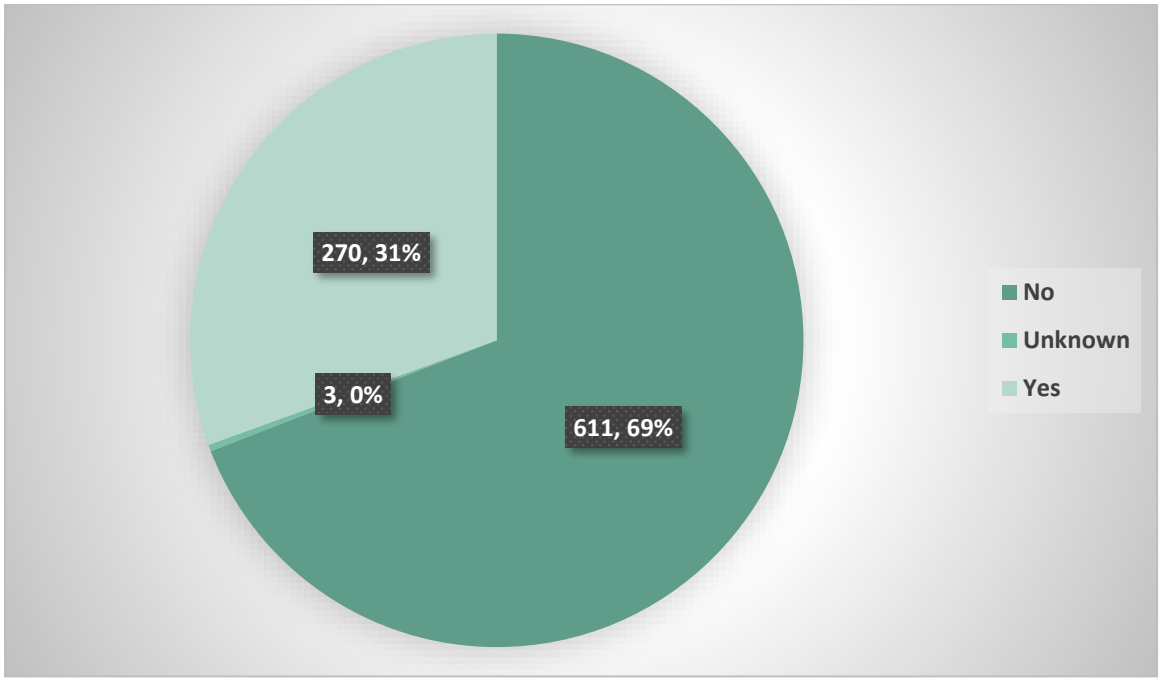
2.1 Recurrent Infection:



Graph 2.1: Recurrent Infection in Primary Immune Deficiency patients (n=884).

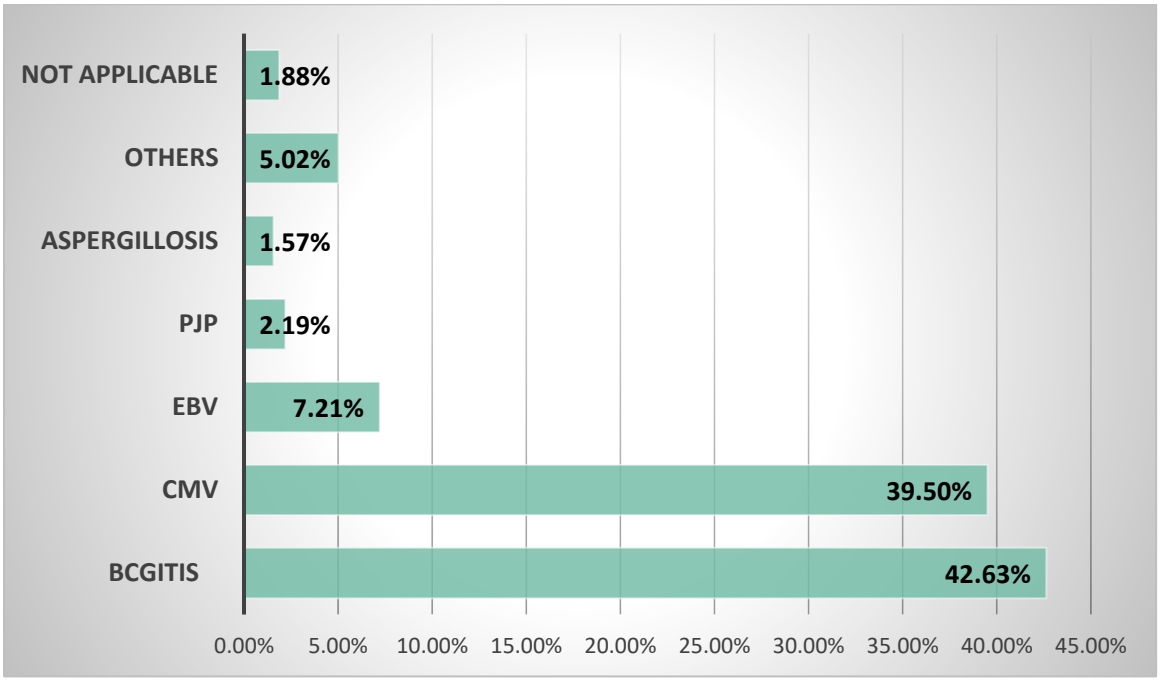
This graph shows that most of PID patients had a recurrent infection by (63%).

2.2 Atypical Infection:



Graph 2.2: Atypical Infection in Primary Immune Deficiency patients (n=884).
This graph illustrate that PID patients by (31%) had atypical infection.

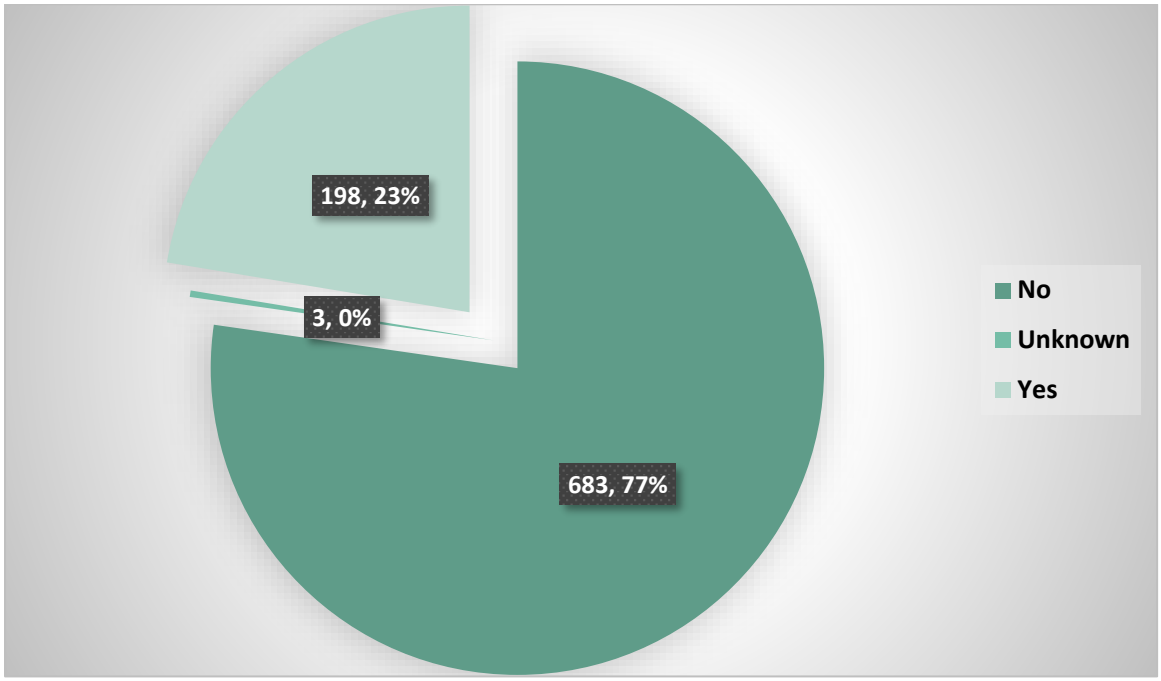
2.3 Subtypes of Atypical Infection:



Graph 2.3: Subtypes of Atypical Infection in PID patients who have a Atypical Infection (n=270).

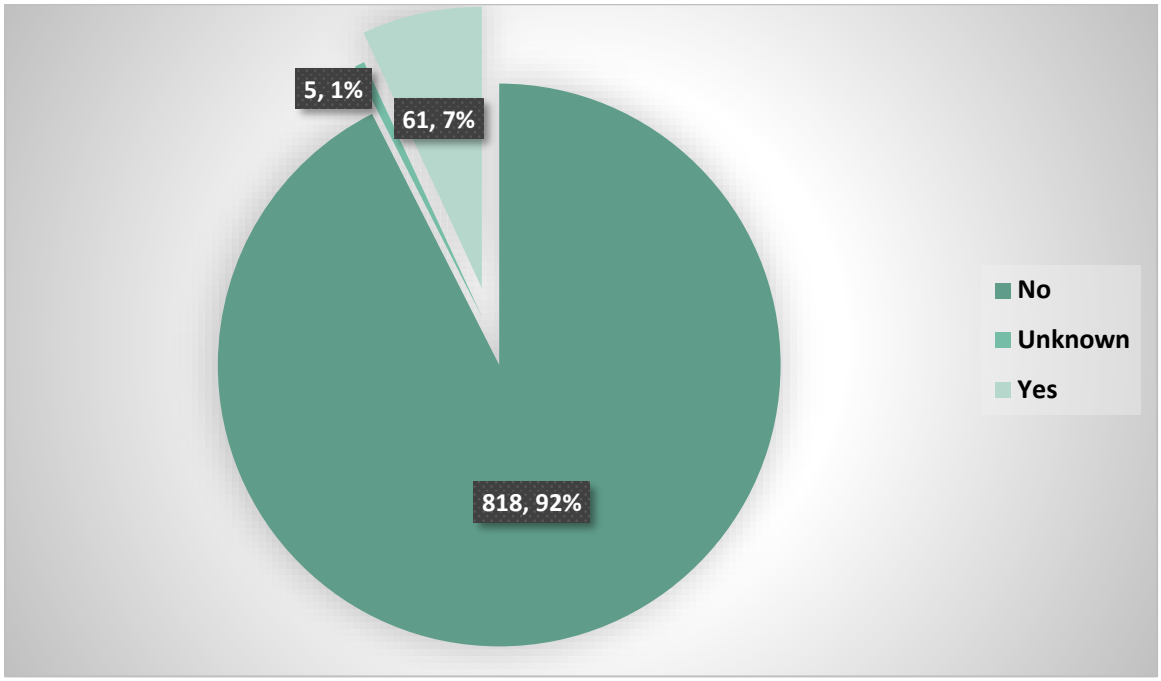
This graph demonstrate that the highest percentage of the Atypical Infection subtypes that was selected is BCGITIS by (42.63%) and the lowest selection was Aspergillosis by (1.57%).

2.4 Failure to Thrive:



Graph 2.4: Primary Immune Deficiency patients with Failure to Thrive(n=884).
This graph shows that the majority of PID patients had no Failure to Thrive (77%).

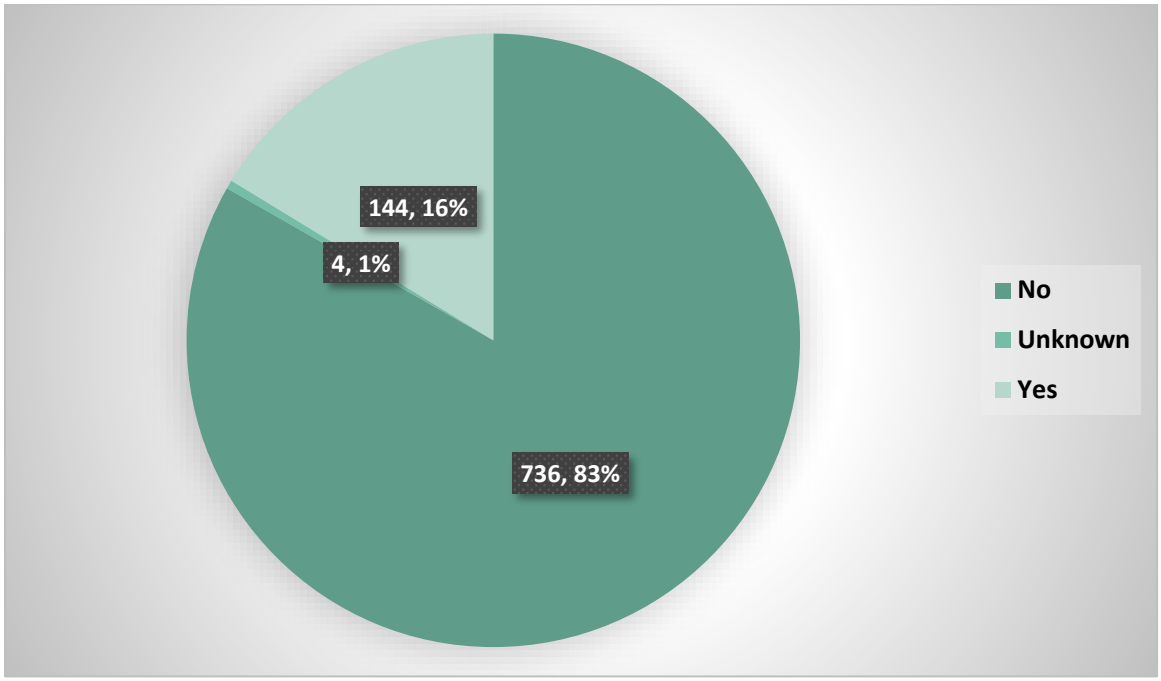
2.5 Recognized clinical syndrome:



Graph 2.5: Primary Immune Deficiency patients with Recognized clinical syndrome(n=884).

This graph demonstrate that most of PID patients had no recognized clinical syndrome (92%).

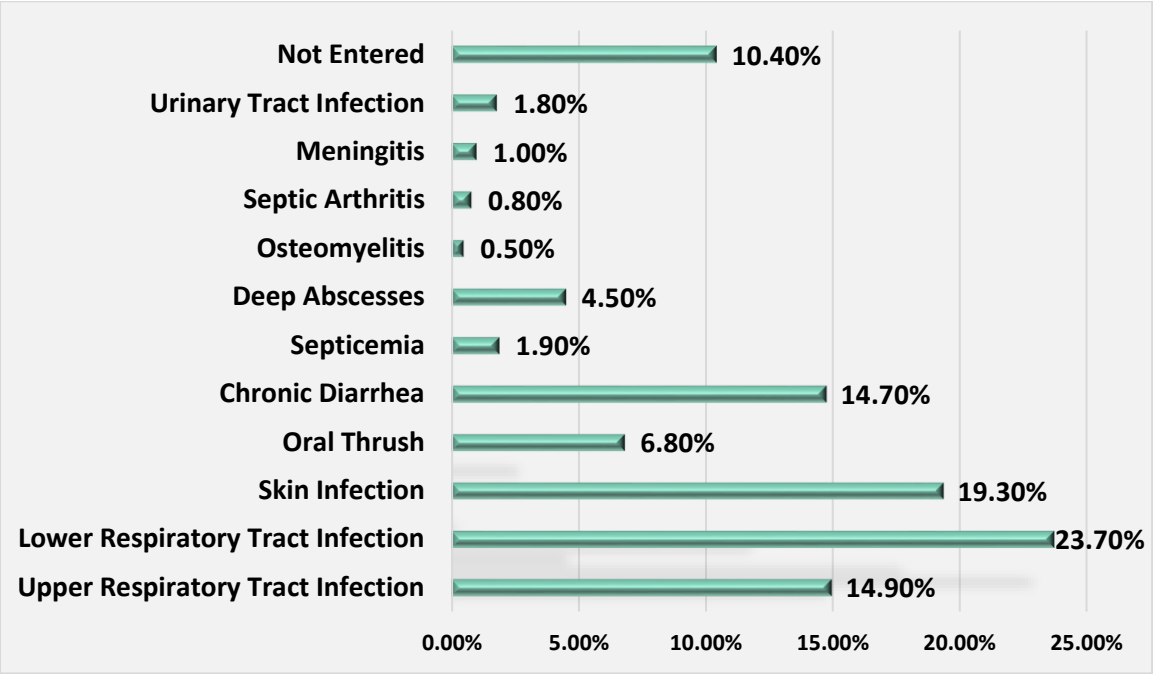
2.6 Target new born screening:



Graph 2.6: Target new born screening in Primary Immune Deficiency patients (n=884).

This graph shows that more than three quarters of PID patients had no target new born screening (83%).

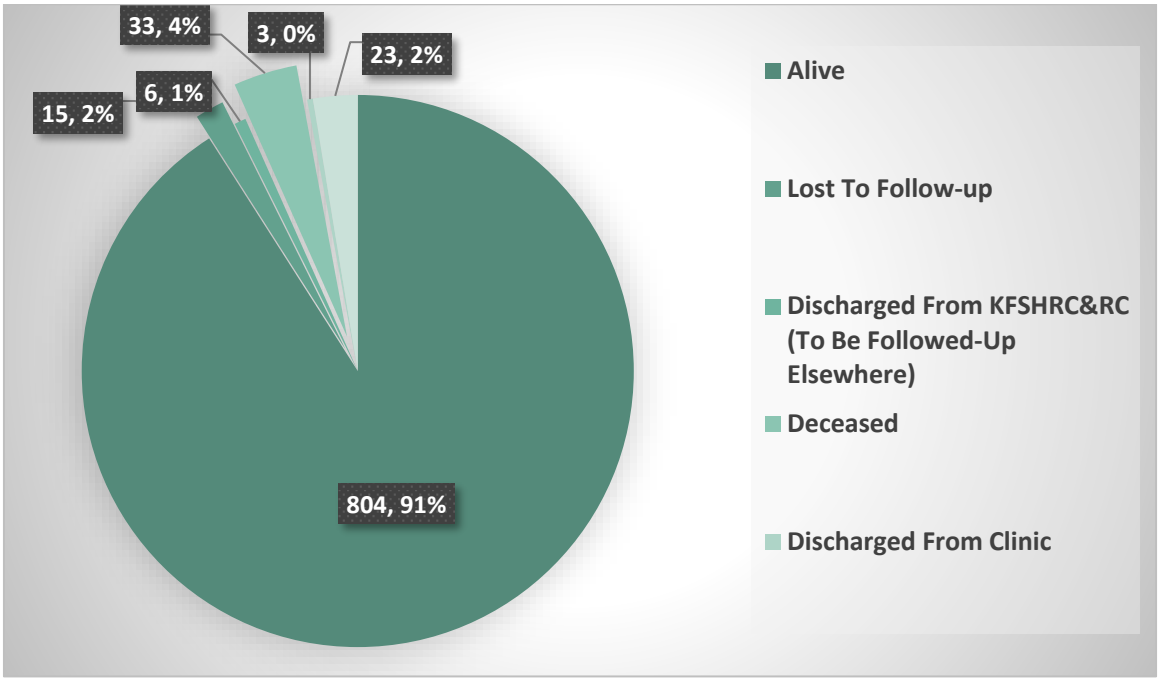
2.7 Type of Infection:



Graph 2.7: Infection type in Primary Immune Deficiency patients (n=884).

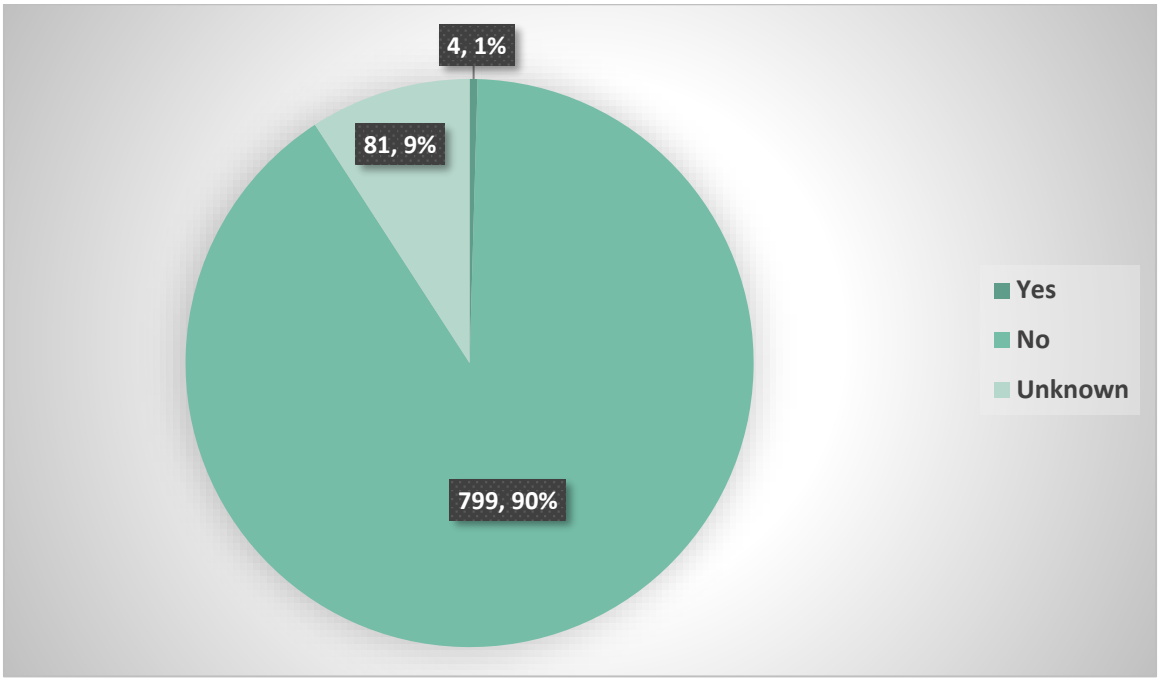
This graph demonstrates that the highest type of infection that was selected was Lower Respiratory Tract Infection (23.7%).

2.8 Current Status:



Graph 2.8: Current Status for Primary Immune Deficiency patients (n=884).
This graph shows that the majority of the patients were alive (91%).

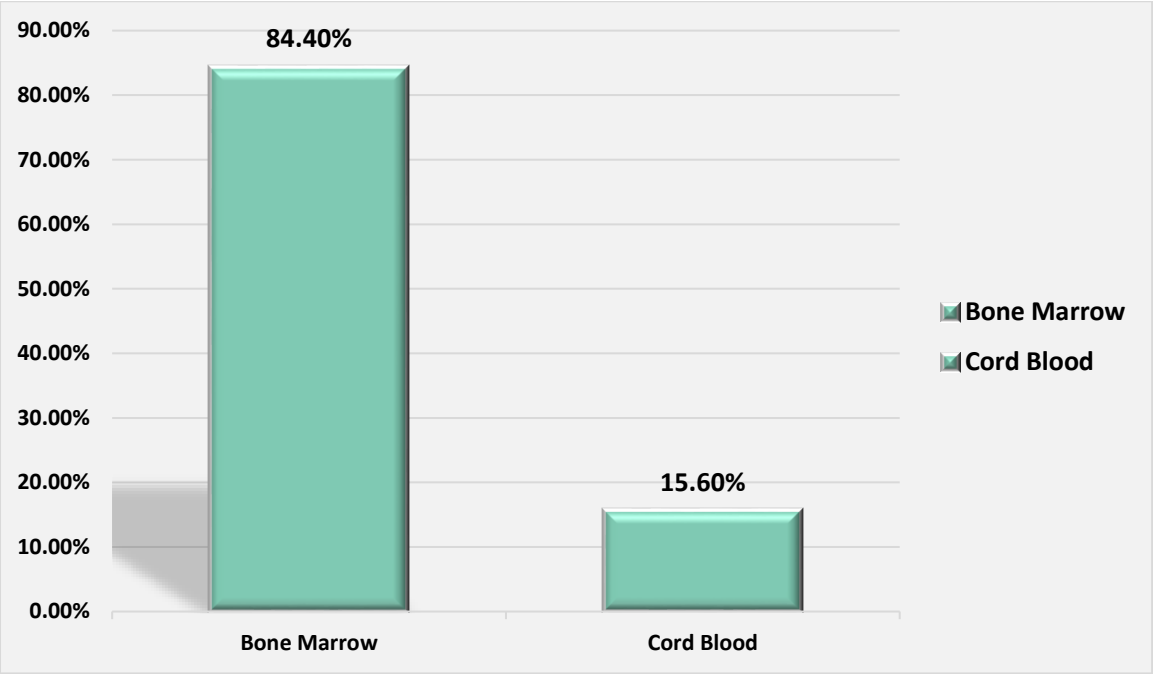
2.9 Smoking:



Graph 2.9: Smoking status in Primary Immune Deficiency patients (n=884).
This graph demonstrates that the majority of the patients were nonsmokers (90%).

3.0 Transplantation

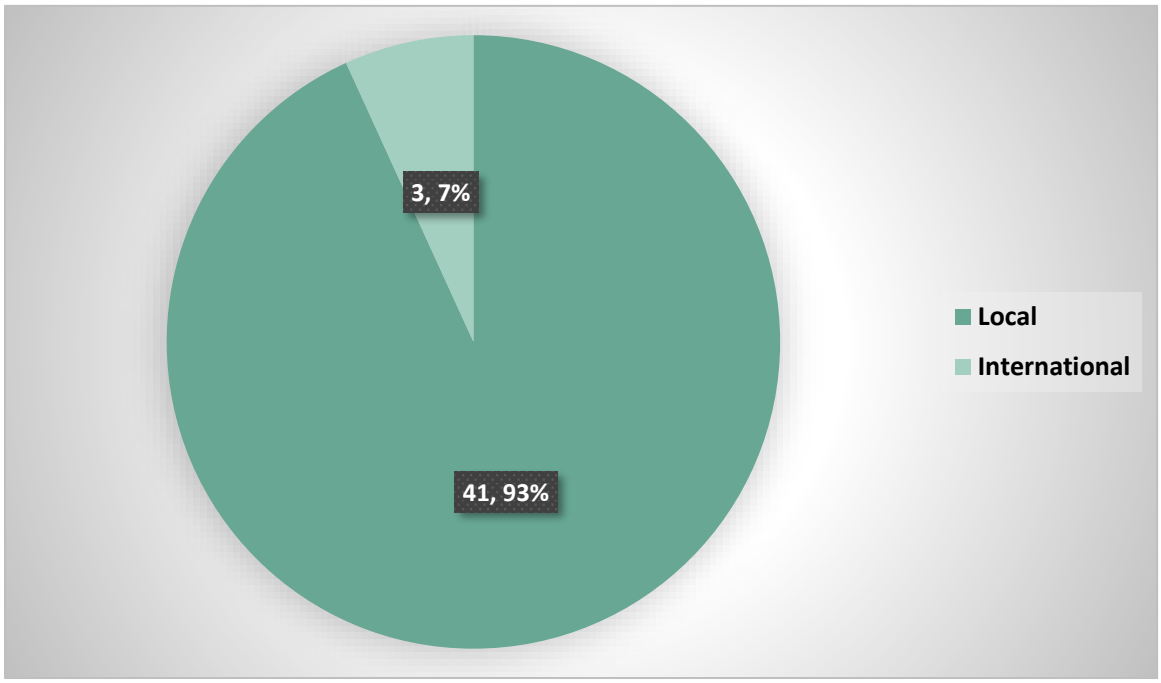
3.1 Transplant Organ in Transplanted PID patients:



Graph 3.1: Type of Transplant organ in Transplanted Primary Immune Deficiency patients (n=288).

This graph shows that most of the patients had a bone marrow transplant (84.40%).

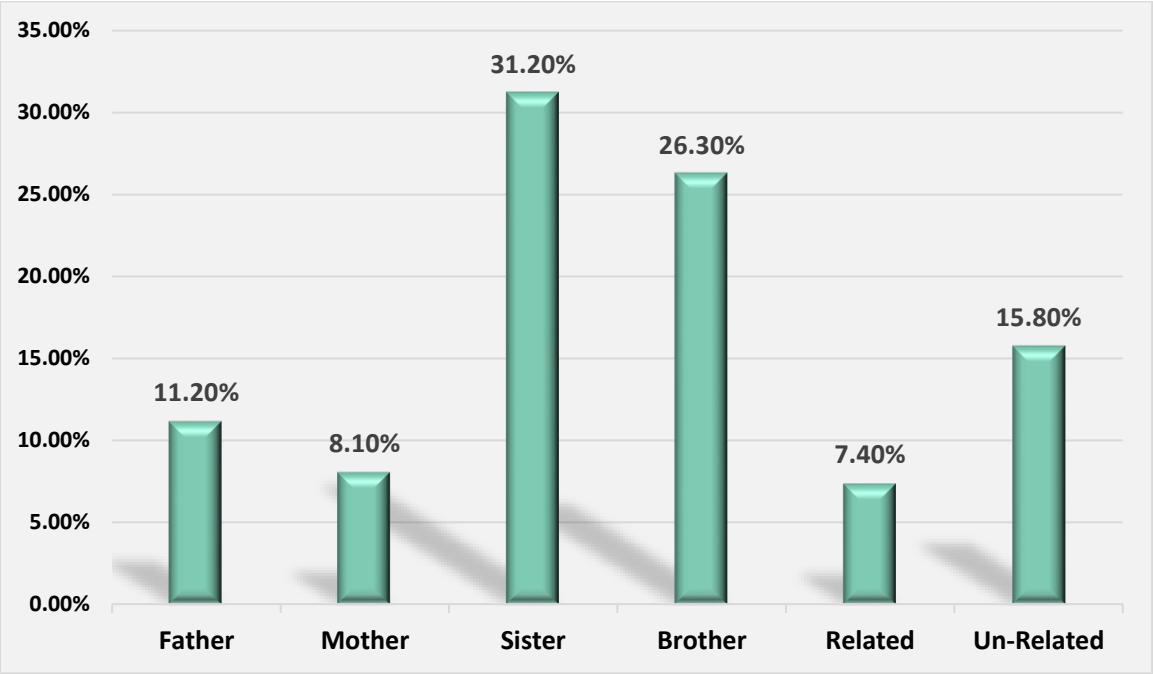
3.2 Place of Transplantation in Cord Blood Transplant Patients:



Graph 3.2: Place of transplantation in cord blood transplant Primary Immune Deficiency patients (n=44).

This graph emphasizes that the majority of blood cord transplantation in PID patients were done in a local Hospital (93%).

3.3 Donors to Transplant PID Patients:

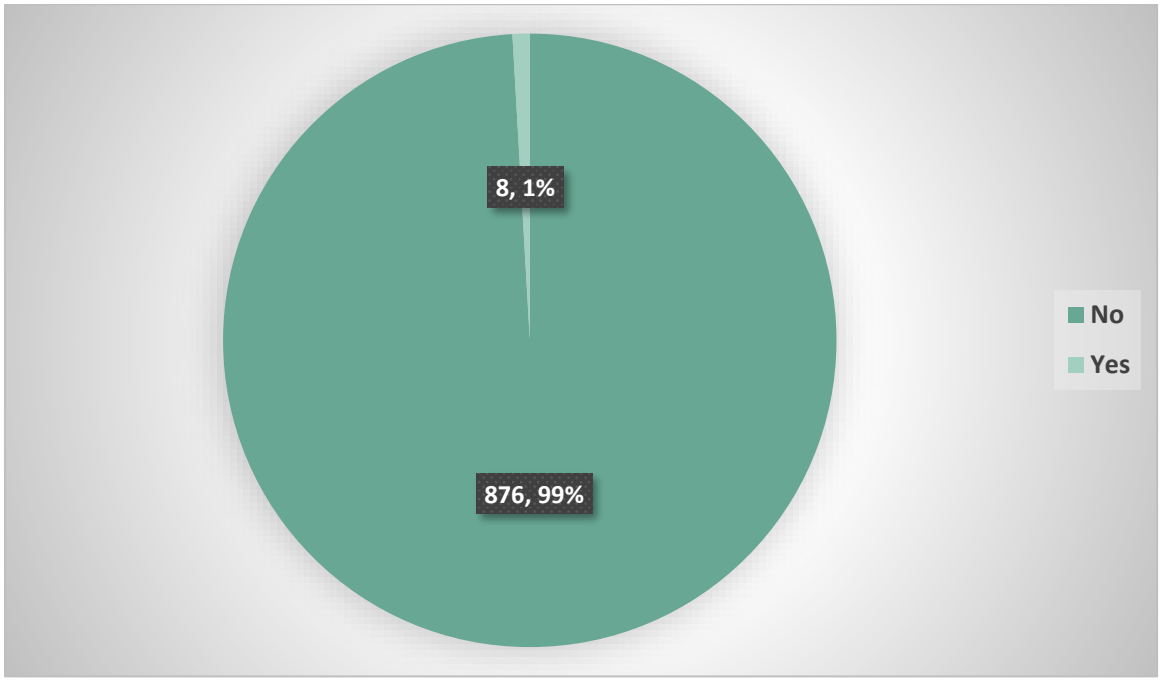


Graph 3.3: Donors Relationship to Transplanted Primary Immune Deficiency patients (n=285).

This graph shows that sisters of the PID patients were the most frequent donors by (31.20%) and related relationships were the less frequent by (7.40%).

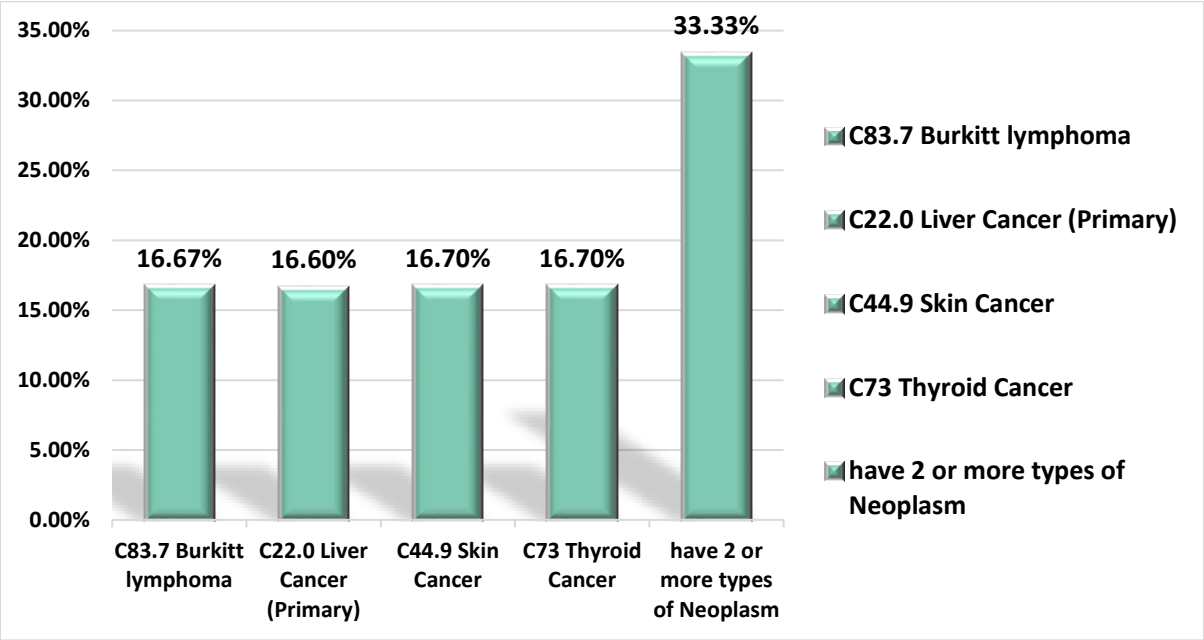
4.0 Concomitant Disease

4.1 Neoplasm:



Graph 4.1: Neoplasm in Primary Immune Deficiency patients (n=884).
This graph highlights that most of PID patients have no neoplasm (99%).

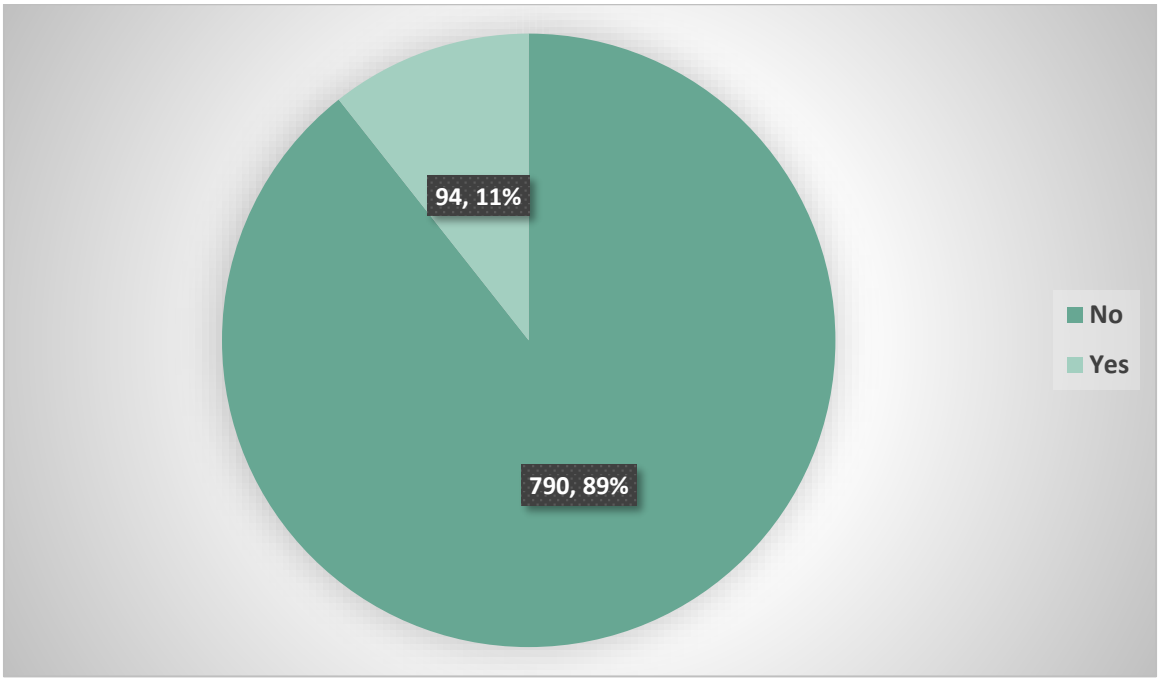
4.2 Neoplasm types:



Graph 4.2: Neoplasm types in Neoplasm Primary Immune Deficiency patients (n=6).

This graph displays that PID patients had 2 or more types of neoplasm by (33.33%).

4.3 Auto-immune:



Graph 4.3: Auto-immune in Primary Immune Deficiency patients (n=884).
This graph shows that the majority of PID patients had no auto-immune (89%).

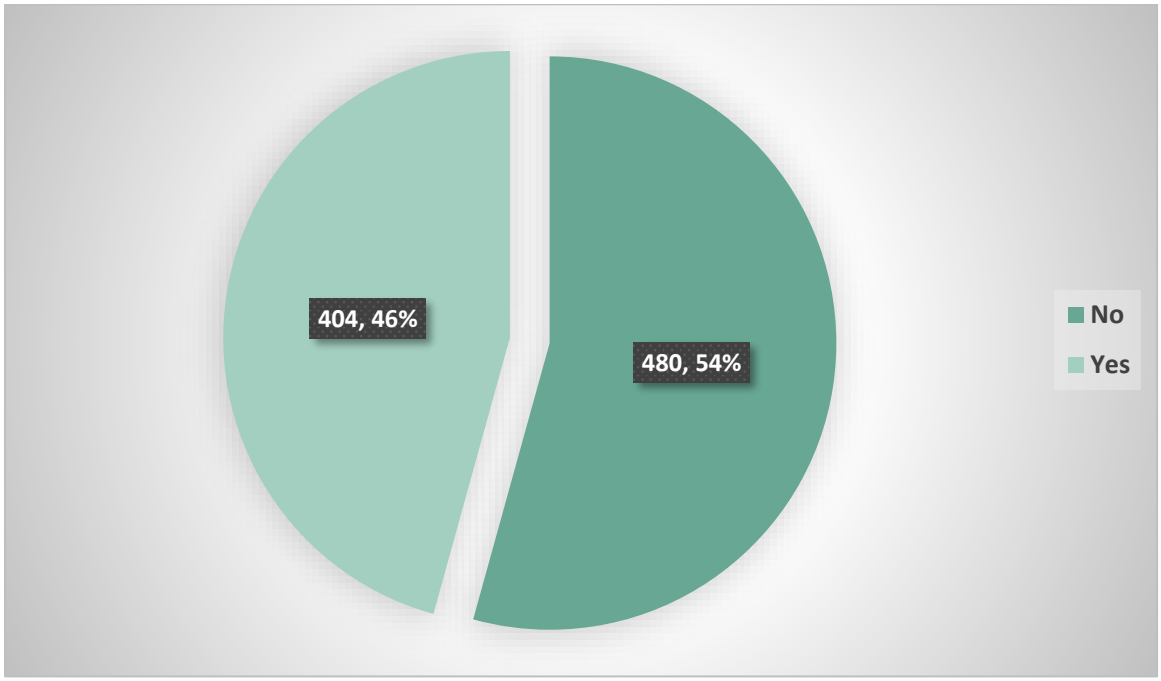
4.4 Auto-Immune Types:

Table 4.1: Auto-Immune Types in Auto-Immune Primary Immune Deficiency patients (n=94).

	Frequency	Percent
L63 Alopecia areata	1	1.1
M13.9 Autoimmune Arthritis	2	2.1
D68.8 Antiphospholipid syndrome	1	1.1
D59.1 Autoimmune hemolytic anemia	13	13.8
K75.4 Autoimmune Hepatitis	1	1.1
D69.3 Idiopathic Thrombocytopenic Purpura (ITP)	1	1.1
E10 Autoimmune type 1 diabetes	1	1.1
G61.0 Autoimmune Guillain-Barré	2	2.1
M35.0 Autoimmune Sjögren's syndrome	1	1.1
M32 Systemic lupus erythematosus (SLE)	1	1.1
L95 Autoimmune Vasculitis	1	1.1
L80 Autoimmune Vitiligo	1	1.1
K90.0 Coeliac Disease	1	1
D69.6 Thrombocytopenia (unspecified)	16	17
L20 Eczema	2	2.1
E03 Hypothyroidism, Other	1	1
K71.0 Toxic liver disease with cholestasis	1	1
Unknown	47	50

As shown in the table the most frequent type of auto immune is Thrombocytopenia (17%).

4.5 Other Concomitant Diseases:



Graph 4.5: Other Concomitant Diseases in Primary Immune Deficiency patients (n=884).

This graph emphasizes that half the patients had other concomitant diseases (54%).

4.6 Types of Other Concomitant Diseases:

Table 4.2: Other concomitant diseases distribution in PID patients who have other concomitant diseases (n=480).

	Frequency	Percent
Bronchial Asthma	60	12.5
Renal failure	3	0.62
Diabetes Mellitus	1	0.2
Hypertension	10	2.1
Stroke	1	0.2
Epilepsy	4	0.83
Abscess	1	0.21
DVT	1	0.2
Hepatitis	3	0.62
thrombocytopenia	1	0.2
Neutropenia	2	0.42
Eczema	19	4
chronic diarrhea	4	0.83
gastritis	1	0.2
Pneumonia	2	0.42
sinusitis	1	0.21
Bronchiectasis	17	3.54
BCGitis	29	6.1
2 or more Other concomitant diseases	219	45.6
Unknown	101	21

This graph highlights that the majority of the PID patients had 2 or more concomitant diseases by (45.6%).

Support Research Will Help

Immunodeficiency Patients

To Develop There Quality of Life.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