

# Past and Future of J Project Meetings in Turkey: Evaluation of Last 10 Years

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## Introduction

Primary immunodeficiency disorders (PIDs) are hereditary inborn errors of immunity characterized with susceptibility to recurrent infections, allergy, malignancy and autoimmune diseases (1). More than four hundred genetic defects caused to PIDs are described up to now and the number is increasing every day. Despite rapid increase of the numbers of identified genetic defects, the awareness of PIDs is not high enough among the physicians, medical students and families (2, 3) which show the need to a PID awareness program. The J Project is a PID awareness, physician education and clinical research collaboration program which established in Eastern and Central Europe (ECE) to increase the knowledge of physicians on PID in 2004 (4).

Turkey has population of more than 80 million people and build a bridge between Europe and Asia. The geographic, economic and cultural characteristics of Turkey are different in the west and east sides of country (5). The frequency of PIDs is not exactly known because PID screening program have not started in Turkey yet. However, the autosomal recessive inheritance diseases have high frequency due to increased ratio of consanguinity in Turkey than in the literature (3, 6, 7). Although consanguineous marriage ratio is 8% in the West, the ratio reach to 50% in the East of Turkey with a mean rate around 21% nationwide. The increased level of consanguinity contributes to the high prevalence of autosomal recessive forms of PID (5). This shows increased need to PID awareness programs in Turkey.

## The J Anatolia Project

Turkey joined with the J Project in 2009 (8) and is continuing to organize J Project events such as J Project Meetings and J Project Congresses by increasing numbers every year (9). The first J Project Meeting was organized in Konya in 2009 and called as the J Anatolia Project (JAP) because Konya, also known as City of Mevlana, is placed in central Anatolia. The rate of consanguineous marriage is around 22% in the city and this ratio reflects the the average consanguinity ratio of Turkey (5). So, it can be estimated that the range and the characteristics of PIDs in Konya will reflect the distribution of PIDs in our country.

Two thousands baby were born for a period of five years and there is only one pediatric immunology center between 2001 and 2006 in Konya. Twenty patients with severe combined immunodeficiency (SCID) were diagnosed in this term in Konya. The incidence of SCID is about approximately 1 per ten thousand live births (6). The average age at diagnosis was 10 months for SCID patients in Konya in this term but the diagnosis age was reported as 4-7 month in the same periods in Europe and USA. When compared to literature diagnostic delay ratio in Konya and in İzmir (10), another city in Turkey, was two to three times higher than in European countries which shows the low awareness on SCID/PIDs between physicians in Konya (5, 6, 10, 11).

We have organized seven J Project meetings in Konya in last 10 years and the number of J Project Meetings are increasing year by year in Turkey (Table 1 and Figure 1). During the last ten years (2009-2019), 47 patients with SCID were diagnosed and the ratio of live birth was around 40.000 babies for each years in these periods in Konya. This results were supporting our previous results and showing that the frequency of SCID is at least 3-4 times higher than European countries. Following the J Project Meetings in Konya and around, the age of diagnosis was decreased to 2 months from 10 months in the last ten years. The ratio of bone marrow transplantation for SCID patients increased

from 28 % to 58 % and the survival rate increased from 20 % to 50 % in Konya in the last ten years. It is not possible to compare ratio of SCID in Turkey with other countries because we have not started a national newborn screening program (NSP) for PID in Turkey yet.

NBS for SCID using a T-cell receptor excision circle (TREC) assay has been established worldwide, including in most of United States, Taiwan, Israel, New Zealand, some Canadian regions, Norway and Spain. The published results showed an incidence of SCID of around 1 in 56,000 newborns in these countries which is two times higher than previously reported ratio (12). Hereby, it would be a realistic approach to expect the ratio of SCID would be more than 4 times higher the other countries in Turkey when we start NBS program in Turkey.

## The East Anatolia J Project

The J Project Meetings were called as the JAP during the first 3 years. The JAP aimed to improve the awareness of PID in physicians and support the early diagnosis and treatment of the patients with PID in Turkey. Because the most immunology centers were mainly located in West Turkey (11), we have just started to organize J Project Meeting in East Turkey where the consanguinity ratio is very high (40%). Thus, the second part of the J Project Meetings were called as the East Anatolia J Project (EAJP). Four J Project Meetings were organized in East Anatolia in Diyarbakır, Şanlıurfa, Malatya and Erzurum between 2012 and 2014 (Table 1 and Figure 1). Following these meetings, several patients were referred us for evaluation of PIDs. This shows importance of J Project awareness meetings to increase the diagnosis of PIDs.

We have organized EJAP Meeting in Van (2017), Batman (2018) and Antakya (2019). We will continue to organize J Project Meetings in Eastern part of Turkey in future.

## J Project Meetings in Western Turkey

We have continued to organize J Project Meet-

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ings in Western part of Turkey such as Sapanca (Sakarya), Antalya, Eskişehir and Konya in 2016 and 2017 for JAP. We also have a good collaboration with University of Medical Sciences and The Society of Family Physicians and organized J Project Meetings in Bodrum (2018), Çeşme (2019), and in Antalya (2019) during their Congresses.

### J Project Congresses

We had hosted three J Project Congresses in Antalya (2014 and 2016) and in Konya (2019). The first J Project Congress (also 100th J Project Meeting) was so important because the PID Declaration was accepted during this meeting and named as Antalya Declaration. The Antalya Declaration for PID called for a prompt and effective action plan to apply current knowledge in diagnosis and treatment of PID patients so that their quality of life would greatly improve. The second J Project Congress (138th J Project Meeting) also was a meeting which shared the recent progress and knowledge in PID field with all the participants. The third J Project Congress (234th J Project Meeting) in Konya was so impressive meeting with its high scientific level and also very important for updating the PID Declaration.

We were also so honored to organize these J Project Congresses to provide a scientific forum to discuss and update recent developments of PID and aim to increase collaborative studies in the field of PIDs especially from Eastern Europe.

### J Cyprus Project

At the end of 2019, a J Project Meeting was organized in Turkish Republic of North Cyprus (TRNC) to discuss the awareness of PID diseases among the physicians and medical students at Near East University Hospital. This was a unique J Project Meeting which was organized in Nicosia with a honorable support by Prof Laszlo Marodi and great collaboration with Prof Nerin Bahçeciler. There are 326,000 local and 150,000 settlers from other countries and 100 pediatri-

cians in TRNC. However, there were only two patients with PID diagnosed until 2010. Following the establishment of a Pediatric Immunology Center in Cyprus, the number of patients diagnosed as PID was increased up to 100 patients with PID in the last ten years. We think that the Cyprus J Project should be continued with additional meetings to increase the awareness of PID among the pediatricians for providing the early diagnosis and treatment of the patients.

### Conclusion

After the first J Project Meeting in 2009, we have organized 25 J Project meetings (Tablo 1 and Figure 1) in the last ten years to increase the awareness of primary immunodeficiency (PID) among physicians for improving the early diagnosis and early treatment. There were about 2500 participants who attended these J Project meetings. The J Project was affiliated with Journal of Pediatric Infection Diseases in 2018 and also with Turkish Clinical Immunology Society in 2019 in Turkey. J Project Meetings in Turkey would allow a valuable and important education for the participants with its unique approach and contribute to increase PID awareness in physicians and early diagnosis and treatment of the patients. The location and the professional background of the PID group in Konya provided the successful management of the J Project Meetings in Turkey.

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