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LETTER TO EDITOR

The J Daughter Siberia Project

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In recent decades, there has been an unprecedented increase in knowledge of the previously neglected field of primary immunodeficiency diseases (PID) in developed countries, particularly in Western Europe and the USA [1]. In sharp contrast, there are still countries with total populations in the tens of millions for which there are no diagnosed and registered PID patients, especially in Eurasia and Africa. Rigorous and coordinated activities by medical authorities, health ministries, and national and international societies and foundations are therefore required to support local teams and institutional groups in launching PID-oriented programs for education and clinical research. The J Project (JP) program was implemented in Central Europe in 2004, to overcome the problem of PID underdiagnosis and undertreatment in most countries in this part of the world [2]. We describe here how the J Siberia Project (JSP) was created and has spread throughout the Urals and Siberia and subsequently to Central Asia to increase awareness of PIDs. As no PID-focused education program existed in the region, a professional team in Ekaterinburg came together to establish the JSP Educational Committee, to design, coordinate, and run the project. Given the complex clinical and immunological phenotypes and genotypes of the diseases already known (more than 200 estimated clinical illnesses and more than 140 PID-causing genes had been discovered by 2009), we included clinical immunologists, research immunologists, and geneticists among the leaders of this project. In particular, geneticists familiar with both gene sequencing and classical cytogenetics and the analysis of chromosomal abnormalities were invited to join us, with the aim of providing strong educational programs on the variable expressivity and incomplete penetrance of a number of monogenic PIDs. Approaching PIDs from different standpoints (the clinical, immunological, biochemical, and genetic standpoints) has been a key concept in our 10 years of educational activity. Review papers presented by local opinion leaders, guest speakers, and practical case presentations have formed part of the programs implemented. Following the general opening of the project, conceptual lectures on the JP and its achievements and the importance of PIDs in modern molecular medicine took place. Particular emphasis was placed on specialist education in hospitals, as we expected most children and adults with suspected PIDs to be referred to hospital clinicians, but family practitioners and general pediatricians have never been discouraged from attending educational events. The JSP meeting series was launched in Ekaterinburg, the capital of the Ural region, with subsequent meetings taking place in the second largest city, Chelyabinsk, also in the Urals, and then in Krasnoyarsk and Novosibirsk in Siberia (Fig. 1). Table 1 and Fig. 2 show the steady increase in the number of patients diagnosed with PIDs. Feedback was ensured by means of a questionnaire, with the correct answers provided. Part of our budget was spent on basic instruments and reagents for education, for the measurement of serum protein concentrations

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Fig. 1 The spread of the J Daughter Siberia Project from Ekaterinburg, Ural Federal District of Russia, to the Siberian Federal District and countries of Central Asia. Green, J Project countries; beige, J Daughter Project countries; stripes, Central Asian countries that are on the merge to join the J Project

and the determination of lymphocyte counts, and to help laboratory experts to become more useful and creative.

The official language of the JP has been, and will remain English, based on a decision taken by the JP Steering Committee. This has never created communication problems at SC meetings. However, English is not currently the dominant foreign language used for communication between medical practitioners and most specialists in the JSP area. We overcame the language barrier, by successfully applying two measures. The first was the bilingual presentation of slideshows in Russian and English. The other was the use of simultaneous interpreters, but this was found to be less effective for the transmission of messages, because of the unique wording of PIDs and because professional interpreters are not widely available. In addition, from a practical standpoint, simultaneous interpretation also increased the already tight budgets for JSP meetings.

Central Asian Extension of the JSP: the JCAP

At the JP meeting in Astana City in 2017, a workshop with delegates from Tajikistan, Hungary, Kazakhstan, Kyrgyzstan, and Uzbekistan was organized, with a view to developing a plan for extending the J Project to the whole of Central Asia (CA). It took a full year to launch the J Project in Uzbekistan, a country with more than 30 million inhabitants and no registered PID patients before 2018. One year after the first meetings in Tashkent and Bukhara, nine new patients with PIDs were diagnosed. In the following year, two more JP meetings were organized, in Tashkent and Samarkand, and a PID working group and a PID-oriented laboratory were set up in Tashkent. In three countries—Kyrgyzstan, Tajikistan, and Turkmenistan—in which no PID patients had ever been diagnosed, in a total population of more than 21 million, it took even more time to break the ice.

The First PID Awareness Meeting in a Remote Mountainous Country: the Kyrgyzstan Experience

Kyrgyzstan is located in the foothills of Pamir, and much of the country is isolated from the capital, creating problems and dangers in transportation, particularly during the winter. The country has limited economic resources and little access by air. The Ekaterinburg team and the leaders of the JSP set out for the 4-day JP meeting on 18 February 2020, on a night flight from Ekaterinburg. The scene was already set, and Skype sessions were organized to allow people from all over the country to participate remotely in the meeting. It was rewarding to see how local physicians and scientists were prepared for this unique event, wanting to find out more and to improve their understanding of the immunology and genetics of PIDs. We hope that these pioneering efforts of a few professional heroes to spread the J Project in Siberia and CA will

Table 1 Primary immunodeficiency (PID) awareness meetings in the Urals, Siberia, and Central Asia over the past 10 years

Year	Venue		Approximate population of the region $\times 10^6$	Number of participants $\times 10^2$	Number of diagnosed patients	Number of PID centers
	Country	City				
2009	Russia	Ekaterinburg	4,3	1,5	163	1
2010	Russia	Chelyabinsk	3,4	1,9	170	1
2011	Russia	Chelyabinsk	3,4	2,1	192	1
2012	Russia	Tyumen	3,7	2,2	208	1
	Russia	Ufa	4,0	2,1	208	1
2013	Russia	Krasnoyarsk	2,8	2,1	307	1
2014	Russia	Ekaterinburg	4,3	5,0	352	1
2015	Russia	Perm	2,6	2,9	372	1
2016	Russia	Kaliningrad	1,0	3,5	409	1
2017	Russia	Chelyabinsk	3,4	3,1	435	1
2018	Russia	Ekaterinburg	4,3	2,1	496	1
	Russia	Ekaterinburg	4,3	1,2	496	1
2019	Uzbekistan	Tashkent	2,3	2,5	3	1
	Uzbekistan	Bukhara	0,2	3,1	3	1
	Uzbekistan	Samarkand	0,5	2,1	3	1
	Russia	Novosibirsk	2,7	4,2	538	1
	Russia	Perm	2,6	2,5	508	1
2020	Kyrgyzstan	Bishkek	0,9	2,9	0	0
	Russia	Ekaterinburg*	4,3	0,4	578	1
	Russia	Ekaterinburg*	4,3	1,2	578	1
	Russia	Ekaterinburg*	4,3	1,8	578	1
	Kyrgyzstan	Osh*	0,9	7,9	0	0

Meetings in 2020 were mostly online (*)

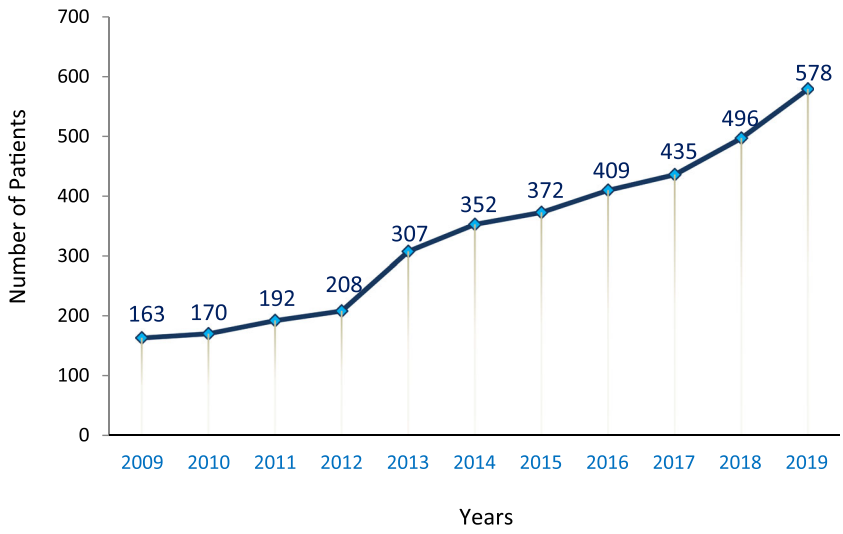

Fig. 2 The number of patients diagnosed with primary immunodeficiencies over the past 10 years in Ekaterinburg, the center of the J Daughter Siberia project

continue and that all the countries of this region will soon be covered by this project.

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Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

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