



## Call to action: Tackling primary immunodeficiencies and cancer

Primary immunodeficiencies (PIDs) are a large and growing group of over 430 rare diseases that occur when certain parts of the immune system are either not present or are not working normally. A compromised immune system can lead to patients being vulnerable to infections, which potentially can be life-threatening, and an increased likelihood of getting a variety of different conditions, including cancer.<sup>1</sup> The risk of PID patients developing cancer can range up to 25% in some PIDs.<sup>2</sup> In fact, cancer is the second highest cause of death (infection being the first) among children and adults with PIDs.<sup>3</sup>

Cancer is one of the main priorities of the EU institutions for a valid reason: in one way or another, these conditions concern us all. Indeed, the European Commission has acknowledged that “*cancer is an individual diagnosis that has important impacts on patients, but also severely affects the lives of their families and friends.*”<sup>4</sup> In the field of PIDs, Common Variable Immunodeficiency (CVID), Ataxia Telangiectasia, Nejmegan-Breakage syndrome, Wiskott–Aldrich syndrome and X-linked lymphoproliferative disease, are just some of the PIDs that have an increased risk of cancer.

After many years of advocacy, the European Commission’s **EU Beating Cancer Plan** and the **Mission Board for Cancer** represent tremendous step in the right direction and offer hope for cancer patients across Europe. These welcomed developments, and the initiatives that will follow, should take high-risk patient populations into account; no matter how rare their conditions are. For this reason and following the IPOPI 17<sup>th</sup> PID Forum titled “*Primary Immunodeficiencies. Fighting Cancer.*”, we at the International Patient Organisation for Primary Immunodeficiencies (IPOPI) are calling on EU institutions to consider the following:

1. *Upcoming cancer screening and diagnosis guidelines need to pay particular attention to high-risk patient populations.*

Early detection offers the best chance of beating cancer, especially for patients with PID. Patients with PIDs should be proactively screened for lymphoma and other cancer types associated with particular PIDs, as certain PIDs have a higher chance of developing certain tumours. Upcoming guidelines for screening and diagnosis targeted towards the Knowledge Centre on Cancer and the National Comprehensive Cancer Centres should acknowledge high-risk patient populations such as patients with CVID, Ataxia Telangiectasia, Wiskott–Aldrich syndrome and X-linked lymphoproliferative disease.<sup>5</sup>

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<sup>1</sup> IPOPI. Primary Immunodeficiencies and cancer (1<sup>st</sup> edition). 2020. Available at: [https://ipopi.org/wp-content/uploads/2021/02/WEB\\_IPOPI\\_PIDandCancer.pdf](https://ipopi.org/wp-content/uploads/2021/02/WEB_IPOPI_PIDandCancer.pdf), last accessed: April 2021

<sup>2</sup> Payam M. and Hassan A. Malignancy in primary immunodeficiency. Available at: <https://www.uptodate.com/contents/malignancy-in-primary-immunodeficiency>, accessed in April 2021

<sup>3</sup> IPOPI, op.cit.

<sup>4</sup> European Commission. Beating Cancer Plan. 2021. Available at: [https://ec.europa.eu/health/sites/health/files/non\\_communicable\\_diseases/docs/eu\\_cancer-plan\\_en.pdf](https://ec.europa.eu/health/sites/health/files/non_communicable_diseases/docs/eu_cancer-plan_en.pdf), accessed in April 2021.

<sup>5</sup> IPOPI, op.cit.

2. *Ensuring continuity of care for patients with multiple conditions should be a specific policy during the formation of multidisciplinary teams.*

In the case of PID patients, if cancer is also detected, a multidisciplinary team comprising an immunologist and an oncologist – or an haematologist depending on the type of cancer involved – needs to ensure that timely and appropriate individualised treatment options are discussed, agreed and started, in the context of the ongoing management of the PID. For this reason, it is imperative to develop and implement a framework for rare disease patients so as to ensure regular communication and continuity of care, including social and psychological support, for PID patients. In the case of cancer patients, and in the context of political discussions on cancer care, cancer related comorbidities need to take a centre part in policy considerations.<sup>6</sup>

3. *Future cancer research programmes goals should consider the relationship between conditions such as PIDs and Cancers*

Studying the relation between PIDs and cancer has the potential to provide insights on the role of the immune system in preventing and/or controlling cancer. The more we understand the biological processes behind PIDs and cancer, the risk factors and health determinants specifically driving cancer, the more effectively we can detect, diagnose, treat and manage them.<sup>7 8</sup> The EU, is in a unique position to coordinate and support research in this area and maximise the potential of studies by pooling scientific expertise, knowledge and real-world data.

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<sup>6</sup> ECPC. Making cancer-related complications and comorbidities and EU health priority. Available at: <https://ecpc.org/wp-content/uploads/2020/07/Final-Cancer-Comorbidities-Joint-statement-1.pdf>, accessed in April 2021.

<sup>7</sup> The European Commission has specifically recognised this for cancer in the Beating Cancer Plan.

<sup>8</sup> F. Hauck et al., Intrinsic and extrinsic causes of malignancies in patients with primary immunodeficiency disorders, available at: <https://pubmed.ncbi.nlm.nih.gov/28669558/>, accessed in April 2021.