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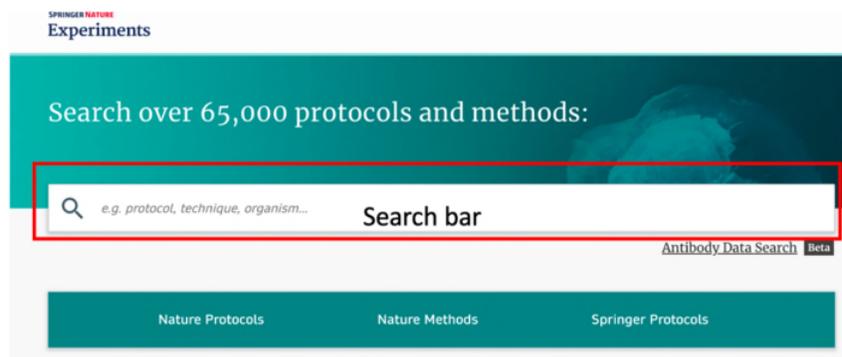
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输入搜索关键词-比如 in vivo imaging mouse。输入“in vivo”后，将显示推荐词条，选择“in vivo imaging”，然后输入“mouse”并单击回车

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- b) 在搜索栏中输入关键字，然后单击回车。示例：流式细胞仪



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Relevance Most recent Most cited Trending

Springer Protocols (2018) Protocol  
 Series: Methods In Molecular Biology > Book: Flow Cytometry Protocols  
**Flow Cytometry Assays in Primary Immunodeficiency Diseases**  
 Maurice R. O'Gorman  
 Inborn errors of immunity are the cause of the primary immunodeficiency diseases, an extremely diverse group of genetic defects that are inherited in Mendelian fashion and result in the impairment of development and/or function of key components of ...more  
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**Models:** Homo sapiens, Capra hircus  
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**Flow Cytometry Assays in Primary Immunodeficiency Diseases**  
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 The primary immunodeficiency diseases (PIDs) encompass an extremely large and diverse number of clinical disorders caused by mutations in genes that affect virtually every measurable component of our immune systems. Many of the genetic mutations lead ...more  
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**Sickle Cell Imaging Flow Cytometry Assay (SIFCA)**  
 Kleber Y. Ferrin, Leigh Samsel, Eduard J. Beers, Laurel Mendelsohn ... J. Philip McCoy  
 Hemoglobin S polymerization under hypoxic conditions in sickle cell disorders causes characteristic shape changes to human red blood cells. Previous sickling assays used to investigate the efficacy of novel agents to treat these disorders are ...more  
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 Series: Methods In Molecular Biology > Book: Human Monoclonal Antibodies  
**One-Tube Multicolor Flow Cytometry Assay (OTMA) for Comprehensive Immunophenotyping of Peripheral Blood**  
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 Recent improvements in the flow cytometry technology allow the determination of the general immune status through the development of multicolor immunofluorescence panels. The one-tube multicolor flow cytometry assay (OTMA) that is presented ...more  
**Techniques:** Immunophenotyping, Flow Cytometry, Multicolor Immunofluorescence  
**Models:** Homo sapiens  
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## Flow Cytometry Assays in Primary Immunodeficiency Diseases

Authors: Maurice R. O’Gorman <sup>1</sup> 1

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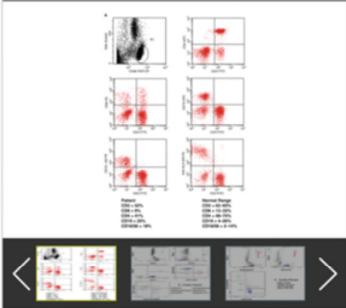
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Abstract 3

Inborn errors of immunity are the cause of the primary immunodeficiency diseases, an extremely diverse group of genetic defects that are inherited in Mendelian fashion and result in the impairment of development and/or function of key components of the immune system. Since the last publication of this chapter in 2011, there have been approximately 100 new primary immunodeficiency diseases officially classified by the “Expert Committee for Primary Immunodeficiency” who met in 2015 and the numbers will continue to rise with the continued evolution and widespread adoption of genomic technologies. The ultimate diagnostic modality involves the identification of a mutation in a gene whose product is known to be involved in immunity. DNA sequencing is however still a rather time-consuming technology. Flow cytometry applications have evolved that are rapid, specific, and relatively inexpensive to screen for abnormalities associated with primary immunodeficiency diseases. The numerous flow cytometry procedures that have been developed to detect abnormalities in peripheral blood cells of primary immunodeficiency patients can barely be covered in an entire book, let alone one chapter. Instead of attempting to cover each disease with a specific assay or test, we will review four procedures each covering one of the three following broad forms of immune abnormalities observed in primary immunodeficiency, i.e., immune subset abnormalities, immune marker abnormalities, and immune function abnormalities. [less](#)

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Fig. 1



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[Immunophenotyping of Human B Lymphocytes in Blood and in Adipose Tissue](#)  
Alain Diaz et al., 2019, Springer Protocols

[Immunophenotyping of Human Innate Lymphoid Cells](#)  
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**Techniques:**  
FACS, Flow Cytometry, Immunophenotyping, Statistical Calculation, DNA Sequencing

**Models:**  
Homo sapiens, Capra hircus

**Others:**  
Lymphocyte subsets, Primary immunodeficiency disease, Oxidative burst, X-linked hyper IgM syndrome (XHIM) CD40 ligand, Autoimmune lymphoproliferative syndrome (ALPS)

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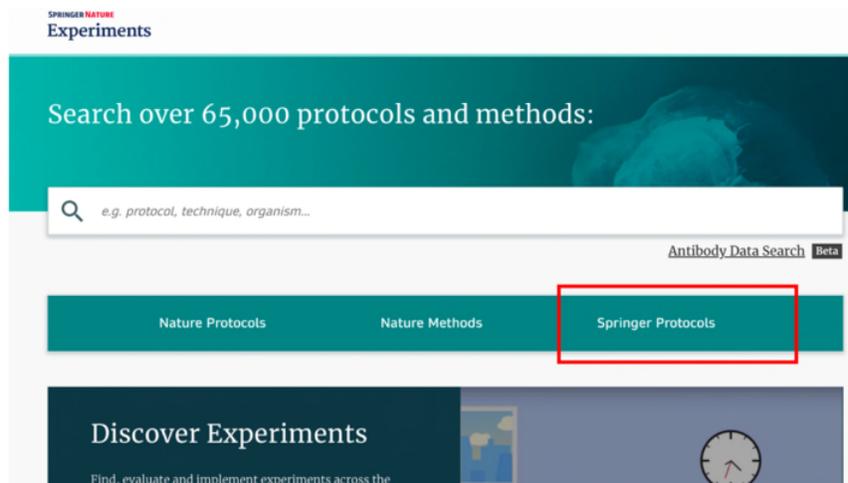
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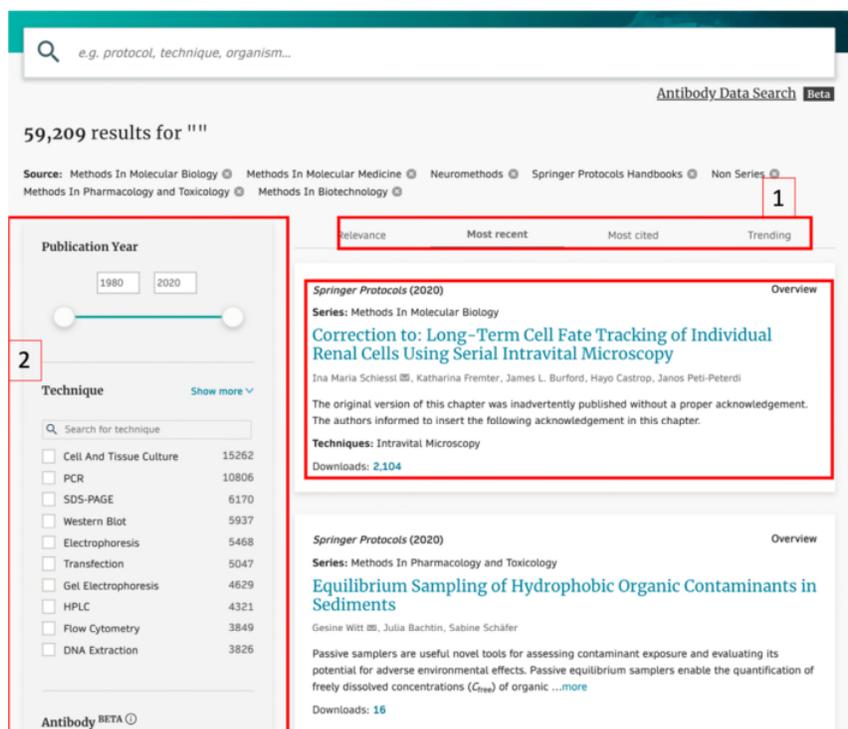
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## Flow Cytometry Assays in Primary Immunodeficiency Diseases

Authors Authors and affiliations

Maurice R. G. O'Gorman

Protocol  
First Online: 26 October 2017

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### Abstract

Inborn errors of immunity are the cause of the primary immunodeficiency diseases, an extremely diverse group of genetic defects that are inherited in Mendelian fashion and result in the impairment of development and/or function of key components of the immune system. Since the last publication of this chapter in 2011, there have been approximately 100 new primary immunodeficiency diseases officially classified by the "Expert Committee for Primary Immunodeficiency" who met in 2015 and the numbers will continue to rise with the continued evolution and widespread adoption of genomic technologies. The ultimate diagnostic modality involves the identification of a mutation in a gene whose product is known to be involved in immunity. DNA sequencing is however still a rather time-consuming technology. Flow

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